Midline one-stage complete unifocalization early outcomes from a single center

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Abstract

Objective: This study aims to present our experience with single-stage complete unifocalization and intraoperative flow study for the repair of ventricular septal defect, pulmonary atresia, and major aortopulmonary collateral arteries.

Methods: This study was conducted through retrospective chart review of all the patients who underwent complete single-stage midline unifocalization in a single tertiary-care institution.

Results: Twenty-two patients underwent midline single-stage unifocalization. The median age was 11 months (IQR: 5–21 months). The number of collateral arteries unifocalized was between one and three (median two). In-hospital mortality was 5%. Follow-up was complete; and the median follow-up regarding survival was 20 months (IQR: 10–28 months). There were three late deaths, and the estimated survival rate was 80% at 10 months and on. Of 22 patients, ventricular septal defect was closed in the first surgery in three patients (14%) and the second surgery in four patients (19%). Total seven patients underwent surgical total repair (32%). Additionally, one out of four patients whose ventricular septal defects were closed with a fenestrated patch is under follow-up with a small ventricular septal defect, while two are waiting for ventricular septal defect closure. Therefore, total eight patients (36%) have reached total correction.

Conclusion: Single-stage unifocalization is a feasible treatment option in ventricular septal defect, pulmonary atresia, and major aortopulmonary collateral arteries. This cohort had unfavorable results regarding the rate of complete repair. The pitfalls encountered were related to problems with meticulous surgical technique, complete unifocalization, and correct implementation of the flow study. (Anatol J Cardiol 2019; 22: 125-31)

Keywords: pulmonary atresia, major aortopulmonary collaterals unifocalization, pulmonary flow study

Introduction

The optimal treatment of ventricular septal defect (VSD), pulmonary atresia (PA), and major aortopulmonary collateral arteries (MAPCAs) is under discussion for the last two decades. Different approaches have been proposed through time, which include: (a) staged unifocalization of the MAPCAs through bilateral thoracotomies, aiming for complete repair at a later stage (1); (b) single-stage unifocalization of the MAPCAs aiming for complete repair at a later stage (2-4); (c) single-stage unifocalization of the MAPCAs aiming for complete repair at the same stage based on the results of an intraoperative flow study (5, 6); and (d) avoiding the unifocalization of MAPCAs; and performing a central shunt between the ascending aorta and the diminutive pulmonary artery, aiming for complete repair after the MAPCAs are eventually closed through catheterization and the native pulmonary arteries grow to a size large enough that a complete repair can be undertaken (7).

We recently initiated a unifocalization program. The method we chose was the single-stage unifocalization with intraoperative flow study. MAPCAs are remnants of the fetal circulation, which normally regress if the flow to the native pulmonary arteries is adequate during the intrauterine life. Otherwise, these collateral vessels become the sole supply to the pulmonary circulation (8). After birth, with the fall of the pulmonary vascular resistance, these collaterals are subjected to increased flow at a higher pressure. This results in development of stenotic segments mainly at two critical anatomic localizations: (a) the join-
ing points of the MAPCAs, which are derived from splanchnic plexus, and the muscular elastic pulmonary arteries; (b) the origins and bifurcations of MAPCAs. One of the main advantages postulated by the proponents of the early single-stage unifocalization is that unifocalization of MAPCAs within the first three to four months of life decreases the time they are subjected to systemic pressures (9). This can be an important reason for higher rate of total correction reported using this method. Additionally, recent reports document complete-repair rates of 73% and 93% using this strategy (5, 6).

This study aims to review our experience with single-stage complete unifocalization and intraoperative flow study for the repair of VSD, PA, and MAPCAs.

**Methods**

This study was conducted through retrospective chart review of all the patients who underwent complete single-stage midline unifocalization in a single tertiary-care institution.

During the study period, unifocalization was performed to patients with VSD, PA, and MAPCA who did not have natural pulmonary arteries or perfused less than 15 lung segments. The patients whose native pulmonary arteries were hypoplastic and supplied at least 15 lung segments underwent a type of systemic-to-pulmonary shunt procedure. These patients are not the subject of this analysis and were not included.

The operations were conducted through midline sternotomy. As far as the hemodynamics of the patients permitted, the MAPCAs were dissected without cardiopulmonary bypass. All the MAPCAs were ligated with the commencement of the cardiopulmonary bypass. Unifocalization was performed on beating heart under cardiopulmonary bypass. Our approach was unifocalization of all the MAPCAs that are not in communication with the native pulmonary arteries (single-supply MAPCAs). The MAPCAs that supplied the lung segments that are also supplied by the native pulmonary arteries (dual-supply MAPCAs) were ligated during the dissection. The techniques used for unifocalization differed on case-by-case basis, with the main purpose being the construction of unobstructed and confluent branch pulmonary arteries and avoiding the exclusion of any lung segments. Once the unifocalization was completed at 28oC, intraoperative flow study was done, using the methodology described by the Stanford group (10) and later modified (11, 12).

The pulmonary artery was cannulated; the lungs were ventilated; and the left atrium was strongly vented. The pulmonary artery pressure was measured, while the flow to the pulmonary arteries was gradually increased to 3 L/m²/min. If the pulmonary artery pressure was higher than 25 mm Hg, a systemic-to-pulmonary shunt was performed. If the pressure was equal to 25 mm Hg or lower, complete repair was undertaken with closure of the VSD and insertion of a right ventricle to pulmonary artery conduit. Right ventricular / left ventricular (RV/LV) pressure rate was measured after CPB in patients undergoing complete repair, and the operation was terminated if the ratio was below or below 0.7. If the ratio was above 0.7, the cardiopulmonary bypass was reinstituted and a 5 mm fenestration was opened on the VSD patch. Algorithm used for intraoperative decision-making is demonstrated in Figure 1.

During the postoperative follow-up, cardiac catheterization was performed for (a) patients who were on extracorporeal membrane oxygenator (ECMO), (b) patients with findings of overflow to the lungs, and (c) patients with persistent low oxygen saturation.

Two types of follow-up were conducted: one based on the latest echocardiographic examination and the other one based on the national death registry. Based on these, two different follow-up periods were calculated and reported.

**Statistical analysis**

Continuous variables were reported as median ± interquartile range (IQR). Categorical variables were reported as n (%). The Kaplan–Meier method was used for survival analysis. The Kaplan–Meier estimated survival time was reported as mean and 95% confidence interval (95% CI). IBM SPSS Statistics Software 21 (SPSS Inc., Chicago, IL, USA) was used for statistical analyses.

**Results**

Twenty-two patients underwent midline single-stage unifocalization. The median age of the population was 11 months (IQR: 5–21 months), and the median weight was 7.8 kg (IQR: 6.5–12 kg).
The male/female ratio was 12/10. On the preoperative cardiac catheterization, 17 patients had confluent pulmonary arteries demonstrated, and five patients had only MAPCAs. The median number of collateral arteries defined on cardiac catheterization was two (IQR: 2–4). Preoperative oxygen saturation was above 85% in 14 patients, and between 75%–85% in eight patients. There were no patients with preoperative oxygen saturation below 75%. Median preoperative oxygen saturation was 86% (IQR: 82%–89%).

The number of collateral arteries unifocalized was between one and three (median: two). There were seven patients whose VSD was closed after unifocalization and flow study. In four of these patients, VSDs were fenestrated due to high postoperative RV/LV pressure ratio. For the remaining 15 patients, who failed on the flow study, some form of systemic-to-pulmonary shunt was performed. Shunt type was Sano shunt in one patient, Blalock-Taussig shunt in 11 patients, and central shunt in three patients.

In the early postoperative period, two patients needed ECMO support. One of these patients could not be weaned off cardiopulmonary bypass due to low oxygen saturation and was put on ECMO in the operating theater. On the cardiac catheterization under ECMO support, there were residual MAPCAs that did not require catheter intervention (Fig. 2). This patient was weaned off ECMO on the postoperative day 2 and had an otherwise uncomplicated postoperative course. The other ECMO patient had a sudden cardiopulmonary arrest on the operation day. This patient was resuscitated through ECMO-CPR. An early cardiac catheterization demonstrated peripheral pulmonary arterial stenoses on the right pulmonary arteries on the anastomotic sites (Fig. 3). Balloon angioplasty was performed for these stenoses. In addition, there was a separate stenosis in the left pulmonary artery that was intervened by placing a stent. This patient developed severe ventricular dysfunction after cardiopulmonary resuscitation that did not improve under ECMO support. On postoperative day 2, the patient died due to progressive multiorgan failure under full flow ECMO support. This patient was the only early postoperative mortality of the population (5%).

In the ICU follow-up, 6 of 19 patients underwent diagnostic cardiac catheterization at median seven days (IQR: 5-20 days). Out of these six patients, two patients had residual MAPCAs, two patients had residual pulmonary stenosis, and two patients had both. In two of four patients with residual MAPCAs, the collateral artery was big enough to cause overflow, so coil occlusion was undertaken. In two of four patients with residual pulmonary stenoses, catheter intervention was undertaken in the same session.

The median postoperative intensive care unit (ICU) stay was 11 days (IQR: 4–30 days) for the 21 patients except for the patient with early mortality. During this period, seven patients (33%) underwent tracheostomy, and two patients (10%) underwent plication of the diaphragm due to phrenic paralysis. One patient underwent a lateral thoracotomy on the 37th postoperative day; and a surgical clip, which was shown to cause bronchial compression on computerized tomography, was removed (Fig. 4). In addition to these, the postoperative course of the patient whose VSD was completely closed during the same session with unifocalization, was complicated. This patient was reoperated on postoperative day 3, due to VSD patch dehiscence and after this operation developed acute renal dysfunction treated with peritoneal dialysis. This patient also had motor axonal neuropathy,
and a tracheostomy was required for weaning from mechanical ventilation. He stayed in the ICU for 63 days postoperatively, and was discharged without tracheostomy on postoperative day 69.

The 21 patients who survived the early postoperative period were discharged on median 21st postoperative days (IQR: 14–60 days); and none of them had any residual organ dysfunction on discharge. Discharge echocardiograms were performed on median postoperative 16 days (IQR: 10–66 days). One patient had moderate and one patient had severe residual pulmonary branch stenosis. Additionally, four patients had residual MAP-CAs demonstrated on echocardiography.

Follow-up was complete; and the median follow-up from the national death registry regarding survival was 20 months (IQR: 10–28 months). There were three late deaths on postoperative months 5, 9, and 10. All three patients had modified BT shunts. The ages of the patients were 1, 4, and 21 months respectively; and none had residual stenoses on their latest follow-up echocardiograms.

On the follow-up, nine patients underwent control angiograms at median 15 months (IQR: 12–21 months). In control catheter angiography findings, pulmonary arborization of two patients was well-developed, and complete repair was performed at 12 and 24
months postoperatively. Another patient who underwent postoperative control angiography was the patient whose VSD was closed (without fenestration) in the first operation. This patient developed infective endocarditis of the RV-PA conduit and anastomotic stenosis on the distal anastomosis. He was reoperated for conduit replacement. All of the other six patients had various levels of pulmonary stenoses; one addressed surgically, another one addressed through catheter intervention; and the other four are on follow-up. Three patients had residual MAPCAs one of which was closed through catheter intervention.

The last echocardiographic follow-up of the 18 early-survivors was on mean 15±11 postoperative months. On the last echocardiogram, one patient had diffuse hypoplasia of bilateral pulmonary arteries, one patient had severe conduit stenosis, one patient had bifurcation stenosis, one patient had severe left pulmonary artery stenosis, and one patient had post-hilar stenosis of the right pulmonary artery.

Out of 22 patients, VSD was closed in the first operation in three patients (14%) and the second operation in four patients (19%). Seven patients underwent surgical complete repair (32%). Additionally, one out of four patients whose VSDs were closed with a fenestrated patch is under follow-up with a small VSD while two are waiting for VSD closure. Therefore, total eight patients (36%) have reached total correction. The treatment pathways of the patients are demonstrated on the flowchart in Figure 5. There were one early and three late deaths, and the Kaplan–Meier analysis estimated survival rate to be 80% at 10 months and on (Fig. 6). The mean estimated survival time was 33 months (95% CI: 27–39 months).

Discussion

Most patients in our cohort underwent a shunt procedure after a failed flow study following unifocalization. In contrast to our series, recent reports by Mainwaring et al. (5) and Carotti et al. (6) had considerably higher rates of single-stage complete repair using this method. Possible reasons for the low rate of single-stage complete repair in our series are problems with surgical technique, incomplete unifocalization, and suboptimal implementation of the flow study.

One of the problems related to surgical technique can be using vessels that are not suitable for unifocalization or anastomotic problems. It is conceivable to assume that this cohort has had a low total correction rate because the pulmonary arteries and MAPCAs were too hypoplastic or had too many distal stenoses at the beginning. Oxygen saturation has been previously used as a guide to estimate the extent of pulmonary artery development in patients with VSD, PA, and MAPCA (5). In this respect, patients with a saturation over 85% can be accepted to have well-developed pulmonary vasculature and are expected to be corrected on the same procedure as the unifocalization. Although this assumption contradicts with the high preoperative oxygen saturation of our cohort, some of the cases can be considered with this respect. An example of such a situation is shown on Figure 2. In this patient, the diminutive pulmonary arteries and MAPCAs were unifocalized, but in the end a hypoplastic neo-pulmonary artery was obtained that later demonstrated suboptimal development. One important lesson we drew from this experience was that MAPCAs with a calibration of 2 mm or less are not good candidates for unifocalization.

It is possible that anastomotic problems caused high pulmonary artery pressure on the flow study. The anastomotic problems can be caused by incomplete dissection of MAPCAs into the parenchyma of the lung. These vessels have tortuous course and have many stenotic segments as they emerge from the aorta to the thorax. Therefore, an important challenge while constructing the unifocalized pulmonary artery is to anastomose these vessels without causing any distortion or impingement by other mediastinal structures and at the same time to extend the anastomosis beyond the stenosed segment. An example of such a situation is demonstrated in Figure 3.

Our results display a high rate of residual MAPCAs on control angiograms. This situation implies incomplete unifocalization that might result in exclusion of certain lung segments from receiving antegrade blood flow. This is in concordance with the fact that pulmonary artery rehabilitation approach reaches lower rates of complete repair with higher RV/LV pressure ratios compared with single-stage unifocalization. In a recent report, Soquet et al. (13) reported a complete-repair rate of 73% with a native pulmonary artery rehabilitation approach, in 33 patients with VSD, PA, and MAPCA. Low RV/LV pressures are also important for the conduit longevity after complete repair. Increased pulmonary artery pressures might lead to earlier conduit dysfunction. A report by Mainwaring et al. (14) demonstrated a negative correlation between increased pulmonary artery pressure and conduit longevity.

Another possible reason of low complete-repair rate is suboptimal implementation of the flow study. In our cohort the pulmonary artery pressure measured on the flow study was either too high to continue with the complete repair or it was misleadingly low so that we initially went to complete repair in five patients and four of them required salvage fenestration of the VSD patch. If the measured pulmonary artery pressure during the flow study is misleadingly low, it is possible to recognize this at the end of the cardiopulmonary bypass. An unexpectedly high RV/LV ratio means that the flow study underestimated the pulmonary artery pressure. On the other hand, if the flow study pulmonary artery pressure is misleadingly high, the patient undergoes a systemic to pulmonary artery shunt operation. After that, it is not possible to know whether the flow study overestimated the pulmonary artery pressure or if the pulmonary vasculature was too immature indeed. The high rate of false-negative flow study outcomes in our cohort implies that the high flow study pressures in some patients might also have been overestimated.
Over the course of this experience, there were certain pitfalls that we learned to avoid while performing the intraoperative flow study. (a) The lungs should be fully ventilated during the flow study. Failure to recruit the atelectatic segments that might have occurred during the unifocalization might lead to overestimated pulmonary artery pressure. (b) The left atrium should be vented, with enough power to aspirate all the flow returning from the pulmonary vasculature to the left atrium. An important sign of inadequate venting is an increase in pulsatility and mean pressure, of the systemic arterial line. Inadequate venting might result in increased left atrial pressure that would lead to overestimation of the flow study results. (c) The flow to the pulmonary arteries should be started low (with 1/5 of the 3 L/min/m² cardiac index) and increased gradually to 2/5 – 3/5 – 4/5 – and 5/5. At each flow, the pressure measurement should be taken after a perfusion of minimum 2 min. Avoidance of these pitfalls is important regarding the false positive results. However, we could not identify any possible risk factors regarding the false-negative pulmonary flow study. Zhu et al. (15) also recently reported that after single-stage unifocalization and complete repair based on pulmonary flow study, 4 out of 40 patients in their cohort required salvage VSD fenestration. Probable reasons for false-negative results for a pulmonary flow study are yet to be delineated.

Early in their experience, many surgeons used morphometric criteria, like total neo-pulmonary artery index, to decide for the closure of the VSD (10, 16). Total neo-pulmonary artery index is sum of the diameters of the MAPCAs and the true pulmonary arteries at the level of pulmonary hilus divided by the body surface area; and it supposedly helps to estimate post-unifocalization RV/LV pressure ratio. Growth of experience on the subject proved that intraoperative flow study is a more sensitive predictor for the tolerance of VSD closure in patients with VSD, PA, and MAPCA (17). Therefore, in the contemporary management of these patients, TNPAI is not required (18). We also did not calculate this index during the preoperative workup of our patients. Overall 33% of patients in our cohort underwent tracheostomy in the early postoperative period. We must admit that after surgery, our threshold for tracheostomy is low. However, airway complications have been previously encountered and reported in midline unifocalization patients. Perri et al. (19) reported four patients who underwent bronchial reconstruction or stenting, out of a cohort of 118 single-stage unifocalizations. In our cohort, the only similar example is the patient who required a thoracotomy to remove the surgical clip that caused external compression. Considering this, early diagnostic workup, including computed tomography and bronchoscopy can be advisable in cases with prolonged mechanical ventilation in the early postoperative period.

It has long been known that the natural history of patients with VSD, PA, and MAPCAs is unfavorable (20). Without intervention, only 10% of these patients can reach the age of 10. Some reports support the palliation of these patients by catheter intervention of MAPCAs (21, 22). However, these reports belong to a time period, before the long-term outcomes of the various surgical approaches to this pathology were available. A recent report by Mainwaring et al. (23) demonstrated a 92% survival rate at a mean follow-up of 5.3 years after midline unifocalization. Based on these outstanding outcomes, we believe that every patient with VSD, PA, and MAPCA should be given the chance of early surgical intervention.

Our results regarding the early and late mortality are comparable to the recent literature (5). In our opinion, after the initial learning curve, this operation can be performed with low perioperative risk. Therefore, early complete unifocalization is a feasible approach, to achieve a healthy pulmonary vascular bed.

It can be discussed that the median age of our cohort (11 months) is rather high compared to the ideal 4–6 month period. This was not our deliberate choice and was mostly related to the delayed admission of the patients. However, it must be noted that high rates of total correction has also been reported in older patient cohorts using the single-stage unifocalization technique (9). Thus, the timing of the procedure should not be restricted with 3–4 months and should be specific to the patient.

**Conclusion**

Single-stage unifocalization is a feasible treatment option in patients with VSD, PA, and MAPCA to supply the pulmonary arteries and MAPCAs with a more physiologic flow. This cohort had unfavorable results regarding the rate of complete repair. The pitfalls we encountered through the course of this experience were related to problems with, meticulous surgical technique, complete unifocalization of MAPCAs, and correct implementation of the flow study. Airway problems can be expected postoperatively and a low index of suspicion is required for early detection of any probable correctable airway problem.

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