



Anesthetic Management during Surgery for Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries

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Background

Tetralogy of Fallot with major aortopulmonary collaterals is a heterogeneous disease with varying degrees of severity requiring complex anesthetic management.

Our institution, Stanford University/Lucile Packard Children's Hospital, has adopted the approach of early complete repair with incorporation of all lung segments, extensive lobar and branch segmental pulmonary artery reconstruction and VSD closure.

We briefly describe our anesthetic approach.

Preoperative

Most critical to the anesthetic assessment is an estimate of overall pulmonary to systemic blood flow.

Patients are categorized as having high, low or balanced pulmonary blood flow. In our series of over 500 patients, half (52.5%) have balanced pulmonary to systemic blood flow (QP:QS=1) while 30.5% have a QP:QS >1 and few, 17.5% have a QP:QS <1.

Patients with either high or low pulmonary flow pose more difficulties with anesthetic management.

Underlying genetic syndromes, such as Alagilles and 22q11 (2.8% and 36% of our patients, respectively) pose additional anesthetic risk.

Pre-Bypass

Approximately, 11% of our patients have concern for abnormal airway anatomy, many of them are 22q11 deletion and will require a pre-repair bronchoscopy.

Though previous surgical repairs were staged procedures, through lateral thoracotomy, our current approach is via median sternotomy, without need for single-lung ventilation.

The majority of our patients are young, 8.6 mos. (4.9; 29 mos.). The number of MAPCAs unifocalized is 3 (2; 5) with cardiopulmonary bypass times of 252 min (191:325). (Median and quartile values reported)

The surgeon's preference is to perform as much of the surgery, as possible, prior to bypass, 262 min (173; 318), to minimize the amount of bleeding when coming off bypass.

Because of the prolonged nature of the repairs, vigilance in blood gas management and need for volume resuscitation in these patients makes reliable arterial and intravenous access a necessity.

In many of the patients, it is a balancing act of managing pulmonary to systemic blood flow.

Thus, It is not uncommon for these patients to become acidotic and hypoxic prior to going on cardiopulmonary bypass, requiring volume administration, mostly blood, and sodium bicarbonate administration.

In some circumstances, the administration of low dose dopamine can help in tenuous hemodynamic states.

Post-Bypass

Because of the amount of dissection and the multiple suture lines for vascular re-anastomoses, post-bypass bleeding can be significant. Our institution has instituted the use of FEIBA (Shire US, Inc. IL; anti-inhibitor coagulant complex) at a dose of 10 units/kg for 1-3 doses.

Based on retrospective analysis, in complete repairs (bilateral unifocalization with VSD closure) achieving a right ventricle: aortic pressure ratio after repair around 0.35 (0.32-0.4) is predictive of improved postoperative outcome.

In our efforts to assist with lowering right ventricular pressure empiric nitric oxide is instituted at 20 ppm.

Conclusion

We have anesthetized over 500 patients at our institution, many (64%) of whom have undergone a single-stage complete repair with VSD closure.

Vigilance in anesthetic management is a key component to the outcomes of these patients and to maintain a low perioperative mortality rate (1.7%).

References

Quinonez ZA, McElhinney D, Hanley F, Wise-Faberowski L et al. **World J Ped Congenital Heart Surg. Accepted for publication 2018.**