

Perioperative Management Of The Child With Tetralogy Of Fallot Undergoing Non-Cardiac Surgery

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Learning Objectives:

After the case discussion, the participants should be able to:

1. List the most common residual cardiac defects following repair of tetralogy of Fallot.
2. Discuss the natural and modified history of tetralogy of Fallot following various repairs.
3. Assess the relevant perioperative issues in the child with repaired tetralogy of Fallot undergoing non-cardiac surgery.
4. List the most appropriate methods/modalities used to assess cardiac function in the patient with repaired tetralogy of Fallot.

Stem Case and Key Questions:

A 16 year old female presents to the preoperative clinic for clearance for an elective cholecystectomy. The patient is a former tetralogy of Fallot who was repaired in childhood. Her mother notes that she had two previous surgeries, and is now completely repaired.

Recently, the patient has had recurrent right upper quadrant pain, diagnosed on ultrasound to be secondary to cholelithiasis and gall stones. The surgeon is planning to undergo a laparoscopic approach.

On physical exam:

Patient is 60 kg, in no apparent distress

HR 75 bpm, BP 110/70, RR 16 and room SaO₂ of 96%.

She has clear lung fields on auscultation, and a loud diastolic murmur.

Key Questions:

1. How often can we expect to be faced with the adolescent or adult with congenital heart disease for non-cardiac surgery?
2. What is tetralogy of Fallot (TOF)?
3. How and when are TOF patients repaired?
4. What is the overall survival and functional status of these repaired patients?
5. What are the long-term complications or sequelae following TOF repair?

6. What is the best modality to evaluate RV function in the repaired TOF patient?
7. What are the indications for reintervention following repair of TOF?
8. Are patients with congenital heart disease (CHD) at increased risk for non-cardiac surgery? Can these patients be risk stratified?
9. Who should do these cases? A pediatric cardiac anesthesiologist, an adult cardiac anesthesiologist, a pediatric anesthesiologist?
10. Should this surgery be done laparoscopically?

Discussion:

1. How often can we expect to be faced with the adolescent or adult with congenital heart disease for non-cardiac surgery?

Current estimates suggest that there are nearly one million adults with congenital heart disease in the United States today and 35,000 babies are born with congenital cardiac defects each year [1]. Only a few decades ago, many congenital cardiac anomalies were fatal in infancy. Now operative repairs are possible in even the most complex anomalies and these infants and children are surviving and growing into adults. Today, approximately 85% of babies born with congenital cardiac anomalies will reach adulthood and long-term survival is expected to continue to improve. The most frequently seen congenital cardiac lesions in the adult population are ASD/VSD, aortic coarctation, tetralogy of Fallot, and transposition of the great arteries [2]. The majority of these adult patients will have undergone one or several surgical or catheter interventions to correct or palliate their cardiac lesion.

2. What is tetralogy of Fallot (TOF)?

Tetralogy of Fallot with pulmonary stenosis represents a spectrum of disease that has in common several anatomic abnormalities; a large non-restrictive VSD, various grades and levels of obstruction to the pulmonary outflow tract, right ventricular hypertrophy, and a rightward deviation of the aortic valve (anterior malalignment of the conal septum) so that it overrides the septal defect. Unrepaired, 75% of TOF patients will die by ten years of age, half of these in the first year of life [3].

3. How and when are TOF patients repaired?

Complete repair of TOF has been performed since the 1950s. Complete correction consists of VSD closure and repair of the RVOTO. Most patients will have had a complete correction either in a single surgery or in two-stages. Acyanotic infants generally undergo primary repair at 3-6 months of age. Cyanotic or symptomatic patients with adequate pulmonary arteries are promptly repaired, irregardless of age. In those infants with diminutive pulmonary arteries a staged repair may be undertaken. Initially, additional pulmonary blood flow is established with an aortopulmonary shunt, an RV to PA conduit, or RVOT reconstruction. The VSD is closed when the pulmonary arterial bed is deemed adequate. Various surgical approaches may be responsible for the varying long-term sequelae. Ventriculotomy

may predispose patients to ventricular arrhythmias. Patch augmentation across the pulmonary valve (transannular patch) results in obligatory pulmonary insufficiency (PI) and leads to RV dilation and dysfunction [4].

4. What is the overall survival and functional status of these repaired patients?

30-year survival rates have been reported at approximately 90%. [5] Most individuals remain in NYHA functional class I or II. Pulmonary insufficiency resulting from transannular patching of the RVOT that may be well tolerated for decades, leads to decreasing exercise tolerance, RV dilation and dysfunction, and an increased risk of sudden death [5-12].

5. What are the long-term complications or sequelae following TOF repair?

- Residual RVOTO
- Residual VSD
- Pulmonary insufficiency (PI)-This is commonly associated with placement of a transannular patch.. The degree of PI is exacerbated by coexisting proximal or distal pulmonary artery stenosis.
- RV dilation-This is most commonly associated with severe and/or prolonged PI.
- RV dysfunction-Many factors may contribute to this. Chronic volume loading due to severe PI, RV dilatation, ischemia and fibrosis, increased wall stress, RV aneurysm or akinesis, or poor myocardial protection during repair are possible contributing factors.
- LV dysfunction-This may be due to inadequate myocardial protection during initial repair, long standing volume overload due to PI, residual VSDs, long-standing systemic to pulmonary shunts, or impairment by altered RV mechanics.
- Aortic regurgitation- This may be due to damage to the aortic valve during VSD closure or dilatation of the aortic root [13].
- Ventricular tachycardia-Sustained ventricular tachycardia is thought to be the underlying cause of sudden death in this population. Risk factors for sudden death include marked RV dilation, QRS duration of > 180msecs, progressive prolongation of the QRS complex, and LV dysfunction [13, 14].
- Atrial tachyarrhythmias-Atrial flutter and atrial fibrillation are most commonly seen and may occur in as many as 1/3 of adult patients.
- Conduction abnormalities-Right bundle branch block (RBBB) is nearly universal in repaired TOF patients. 15% of patients will have RBBB and a left anterior hemiblock but this does not predict complete heart block. Complete heart block rarely occurs in the late postoperative period but can occur in the immediate perioperative period and is secondary to damage to the AV node during VSD repair. A small percentage will require permanent pacing postoperatively.

6. What is the best modality to evaluate RV function in the repaired TOF patient?

It has become increasingly clear that RV dysfunction and failure is the single most important morbidity associated with TOF repair. Pulmonary insufficiency, once thought to be relatively benign, is the principle driving force for progressive right ventricular dilation, declining exercise tolerance, arrhythmias, and sudden death [5-12, 17]. There is substantial evidence that pulmonary valve replacement ‘remodels’ the dilated RV if performed before RV size and function are markedly abnormal [5-7,12-17]. Although echocardiography and magnetic resonance imaging (MRI) are complimentary modalities in assessing PI and RV form and function, MRI has become the preferred method for longitudinally following patients with repaired TOF [18-20]. The optimal timing of pulmonary valve replacement is less certain. Several studies have suggested that earlier PV insertion (RV end-diastolic volume $\sim 150 \text{ m}^3/\text{m}^2$) may lead to normalization of RV volumes and preserved RV function [17-19].

7. What are the indications for reintervention following repair of TOF?

Reintervention is indicated in about 10% of patients after TOF repair over a 20-year follow-up [5, 6]. The following conditions warrant consideration for reintervention:

- Free PI associated with progressive RV enlargement,
- A residual VSD with a significant left-to right shunt.
- Residual patent systemic to pulmonary arterial shunts leading to LV volume overload
- Residual RVOTO or RV-PA conduit stenosis with RV pressures $>2/3$ systemic pressure
- Branch pulmonary artery stenosis, particularly in the face of severe PI.
- Significant aortic regurgitation
- Clinically significant arrhythmias

8. Are patients with congenital heart disease (CHD) at increased risk for non-cardiac surgery?

It is difficult to quantify risk in this population of patients. In a retrospective study reviewing charts of 276 patients under 50 years of age, Warner and colleagues at the Mayo Clinic found current treatment of congestive heart failure, the presence of cyanosis, age younger than 2 years, pulmonary hypertension, inpatient disposition, higher ASA status, and surgery on the respiratory and nervous systems to be associated with an increased risk of perioperative complications. Children with one or more of these risk factors had a complication rate of 11.7% versus 1.7% in children without these variables[21].

Investigators at the University of Michigan found an ASA score of class IV or higher, younger age, pattern of referral (transfer from tertiary center versus outpatient), and emergency operations to be risk factors for 30-day mortality in the *pediatric* population with CHD undergoing general surgery. The complexity of the CHD or the surgical procedure did not seem to influence mortality[22].

Investigators at the University of Virginia queried the University Hospital Consortium database (> 60 university hospitals in the US) for 1, 2, 3, and 30-day

mortality rates in pediatric inpatients with CHD versus those without CHD. Overall mortality was 6.0% in patients with CHD and 3.8% in those without CHD. Age (< 1 yr) and complexity of the underlying CHD were associated with higher mortality rates [23]

9. Who should do these cases? A pediatric cardiac anesthesiologist, an adult cardiac anesthesiologist, a pediatric anesthesiologist?

In 2001, the American College of Cardiology published their recommendations regarding health care for adults with congenital heart disease [1, 24-25]. Adults with moderate and complex CHD who require non-cardiac surgery have special needs to be addressed by the surgical and anesthesia team. Ideally, operations in patients with complex CHD should be performed at a regional adult congenital heart disease (ACHD) center with physicians experienced in the care of these individuals and with the consultation of cardiologists trained in this discipline.

Who should provide the anesthetic care of patients with CHD undergoing non-cardiac surgery [26, 27] ? There is no data to support any position in this matter. In fact, we have yet to define “adequate” training for the pediatric cardiac anesthesiologist. Perhaps more important than the specific training details of those caring for children, adolescent, and adult patients with CHD is the access to expertise and the focus and dedication to the multidisciplinary care of these patients.

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Table 1. Types of adult patients with CHD of great complexity
Conduits, valved or unvalved
Cyanotic congenital heart
Double-outlet ventricle
Eisenmenger syndrome
Fontan procedure
Mitral atresia
Single ventricle (also called double inlet or outlet, common or primitive)
Pulmonary atresia
Pulmonary vascular obstructive diseases
Transposition of the great arteries
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of AV or VA connection not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Table 2. Types of adult patients with CHD of moderate complexity
Aorto-left ventricular fistulae
Anomalous pulmonary venous drainage
AV canal defects
Coarctation of the aorta
Ebstein's anomaly
Infundibular RVOTO
Ostium primum ASD
PDA (not closed)
Pulmonary valve regurgitation (moderate to severe)
Pulmonic valve stenosis (moderate to severe)
Sinus of Valsalva fistula/aneurysm
Sinus venosus ASD
Subvalvar or supra-valvar AS

Tetralogy of Fallot

VSD with absent valves, aortic regurgitation, coarctation of the aorta, mitral disease, RVOTO, straddling TV/MV, subaortic stenosis
