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Original Article

Modern surgical management of patients with tetralogy of Fallot*

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Abstract Tetralogy of Fallot is the most common cyanotic heart defect seen in children beyond infancy. There is considerable variation in surgical management today ranging from differences in timing of complete repair versus initial use of a shunt for palliation, and methods for right ventricular outflow tract reconstruction. This article will examine some of the current surgical management techniques for patients with Tetralogy of Fallot.

Keywords: Tetralogy of Fallot; surgical management

Tetralogy of Fallot is the most common cyanotic heart defect seen in children beyond infancy. Tetralogy of Fallot is also the most common cyanotic congenital lesion that is likely to result in survival to adulthood and thus is the most common complex lesion to be encountered in the adult population after repair.

The original anatomic description of tetralogy of Fallot included a tetrad of malformations:

- ventricular septal defect,
- right ventricular outflow tract obstruction,
- aorta overriding the ventricular septum, and
- right ventricular hypertrophy.

A uniform aetiology may explain this anatomic tetrad. The monology of anterior deviation of the infundibular septum causes hypoplasia of the subpulmonary infundibulum and thus accounts for all components of the tetrad.

Nomenclature and classification

The Definitions Working Group of The International Society for Nomenclature of Paediatric and Congenital Heart Disease (www.ipccc.net) has proposed the following definition for tetralogy of Fallot:[1]

> Tetralogy of Fallot is defined as a group of malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Hearts with tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override; hearts with tetralogy of Fallot will most often have right ventricular hypertrophy.

The four subtypes of tetralogy of Fallot are:

- Tetralogy of Fallot, pulmonary stenosis
- Tetralogy of Fallot, atrioventricular septal defect
- Tetralogy of Fallot, absent pulmonary valve
- Tetralogy of Fallot, pulmonary atresia.

This article will focus on the diagnosis and treatment of the common form of tetralogy of Fallot, which is tetralogy of Fallot with pulmonary stenosis. The pulmonary stenosis may be at the subvalvar, valvar, or supravalvar level.

Diagnosis

Clinical presentation of tetralogy of Fallot is dependent on the degree of obstruction of the right ventricular outflow tract. When obstruction is severe at birth, presentation is in the neonatal period. Severe cyanosis is the most obvious symptom with

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dependency of a patent ductus arteriosus in the most severe cases.

History

Most children with tetralogy of Fallot are asymptomatic at birth. They initially present when a systolic murmur is detected within the first few weeks of life. Development of cyanosis occurs with increasing obstruction of the right ventricular outflow tract and in some cases may be delayed until late childhood if obstruction is not severe. Historically, these children were often seen squatting – to increase their systemic vascular resistance and therefore decrease the amount of shunting away from their pulmonary circulation.

Hypercyanotic attacks or "Tet spells" is an important feature of these patients, which may be caused by infundibular spasm. The episodes result in sudden and dramatic falls in oxygen saturation along with the development of metabolic acidosis. Hypercyanotic attacks are potentially dangerous as they may lead to cerebral damage or even death.

Children with mild levels of obstruction may have features indistinguishable from those with a large ventricular septal defect and appear breathless or feed poorly with poor weight gain.

Physical examination

Essential findings in the neonate with severe tetralogy of Fallot are cyanosis and, on auscultation, a single second heart sound. A systolic murmur may or may not be present. Clubbing of the fingers and toes may be detected at 2–3 months of age. Patients with mild levels of obstruction may only present with a murmur.

Echocardiography

Echocardiography is usually all that is necessary to define the anatomic features consistent with tetralogy of Fallot. Severity of right ventricular outflow tract obstruction is important to distinguish; however, indications for surgery are related more to the presence of symptoms than to the degree of right ventricular outflow tract obstruction demonstrated by echo. It is helpful to determine the presence of an anomalous anterior descending coronary from the right coronary artery across the infundibulum of the right ventricle, but it is not essential information as this will be readily apparent to the surgeon at the time of operation.

Catheterisation

Cardiac catheterisation is rarely needed in the diagnosis of tetralogy of Fallot. Catheterisation may be indicated when the presence of important aorto-pulmonary collaterals is in doubt or questions persist about the anatomy of the pulmonary arterial system or presence of multiple muscular ventricular septal defects.

Treatment

Many of the basic strategies for cardiac surgery in general were developed by visionary surgical pioneers four and five decades ago while performing surgical procedures in, and caring for, patients with tetralogy of Fallot.[2] This article will summarise the current surgical approaches for palliation and repair of tetralogy of Fallot.

Optimal surgical treatment remains controversial. Despite the potential advantages for elective one-stage repair, its use in early infancy is not universally accepted.[3] Some centres prefer a two-stage approach involving initial placement of a systemic to pulmonary artery shunt.[4] Our current approach is for complete repair at diagnosis, unless pulmonary atresia is present. If pulmonary atresia is present necessitating the use of a right ventricular to pulmonary artery conduit, we will initially palliate with a systemic to pulmonary artery shunt to allow for growth and potentially a larger conduit at the time of complete repair. Advantages of a transatrial approach versus a transventricular approach also remain unclear.

Palliative treatment

The initial placement of a systemic to pulmonary artery shunt is indicated in symptomatic neonates with anatomy that is not favourable to corrective repair. This generally refers to neonates who will require placement of a pulmonary homograft to relieve right ventricular outflow tract obstruction. Examples of these conditions include patients with an anomalous anterior descending coronary artery across the right ventricular infundibulum or patients with certain forms of pulmonary atresia.

Initial placement of a systemic to pulmonary artery shunt is also indicated in centres that prefer a two-stage approach for symptomatic neonates. Elective complete repair is usually performed at 4–6 months of age for patients previously treated with shunt palliation.

Placement of a systemic to pulmonary artery shunt is usually performed through a median sternotomy. Cardiopulmonary bypass is not routinely necessary. A modified Blalock–Taussig shunt is used for most patients if necessary. If the patient has a right-sided aortic arch, a central shunt would be preferred.

Corrective treatment

Complete repair of tetralogy of Fallot is indicated in symptomatic neonates or infants. All patients with

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tetralogy of Fallot will require surgical correction. Timing of surgery is based on the presence of symptoms. Elective repair is performed at 3–6 months of age in asymptomatic children.

Corrective surgery is performed through a median sternotomy incision with cardiopulmonary bypass and mild hypothermia (32–34°C). The heart may be vented through the right superior pulmonary vein or atrial septal defect. After aortic cross-clamping, a standard right atriotomy is performed. If a transatrial approach is planned, relief of right ventricular outflow tract obstruction is performed first. The infundibular obstruction is assessed through the tricuspid valve, and parietal and septal bands are divided and resected up to the pulmonary valve annulus until obstruction is relieved. The pulmonary valve annulus is then sized and consideration for a valve-sparing procedure made if the valve annulus is measured at a z-score of at least −2. A vertically oriented arteriotomy in the main pulmonary artery can be used to facilitate examination of the pulmonary valve leaflets and annulus. Condition of the valve leaflets, abnormal thickening, and fusion of commissures should be noted. Division of fused commissures may enlarge the effective orifice to an acceptable size; however, grossly thickened and abnormal leaflets should probably not be saved and a transannular patch performed.

The ventricular septal defect is closed through the tricuspid valve with a prosthetic patch. Gluteraldehyde-treated pericardium is used to repair the pulmonary arteriotomy or for a transannular patch if necessary. A patent foramen ovale may be left open if there is concern about right ventricular compliance following repair. In some centres, the surgical approach for both the relief of the infundibular obstruction and ventricular septal defect closure may be through a ventriculotomy. A ventriculotomy is also necessary when the right ventricular outflow tract is hypoplastic as resection of infundibular muscle bundles will not adequately relieve the obstruction.[5]

Once the repair is completed and the child weaned off cardiopulmonary bypass, a trans-oesophageal echocardiography is used to check the repair. If the pulmonary valve annulus was preserved, the residual gradient across the right ventricular outflow tract is assessed. A gradient of 20–25 mmHg is probably ideal. If right ventricular pressures are measured at >2/3 systemic, a transannular patch should be performed.[6]

Outcomes

The Society of Thoracic Surgeons Congenital Heart Surgery Database was recently queried to ascertain current trends in the management of tetralogy of Fallot and to determine the prevalence of various surgical techniques.[7] The study population (n = 3059 operations) was all index operations in 2002–2007, age 0–18 years with primary diagnosis = tetralogy of Fallot, and primary procedure = tetralogy of Fallot repair or palliation. Patients with pulmonary atresia, absent pulmonary valve, and atrioventricular canal were excluded. The findings of this analysis demonstrate current trends in management and outcomes for surgery in patients with tetralogy of Fallot in the United States of America:

- 294 patients had initial palliation, including 178 neonates.
- 2534 patients had repair of tetralogy of Fallot as the initial operation, primary repair, including 154 neonates.
- 217 patients had repair of tetralogy of Fallot after prior palliation.
- Of patients who had primary repair (n = 2534), 975 had repair at 3–6 months, 614 at 6 months to 1 year, 492 at 1–3 months, and 154 at 0–30 days.
- Of patients who had repair following prior palliation (n = 217), 65 had repair in the first 6 months of life, 111 at 6 months to 1 year, and only 41 (18.9%) at more than 1 year of age.
- Of 2534 primary repairs, 581 (23%) had no ventriculotomy, 571 (23%) had non-transannular patch, 1329 (52%) had transannular patch, and 53 (2%) had right ventricle to pulmonary artery conduits.
- Of repairs after prior palliation (n = 217), 20 (9%) had no ventriculotomy, 30 (14%) had non-transannular patch, 144 (66%) had transannular patch, and 24 (11%) had conduits.
- Discharge mortality (95% confidence interval) was
  - 22 of 294 (7.5%; confidence interval: 4.7–11.1%) for initial palliation,
  - 33 of 2534 (1.3%; confidence interval: 0.9–1.8%) for primary repair, and
  - 2 of 217 (0.9%; confidence interval: 0.1–3.3%) for secondary repair.
- For neonates, discharge mortality was
  - 11 of 178 (6.2%; confidence interval: 3.1–10.8%) for palliation and
  - 12 of 154 (7.8%; confidence interval: 4.1–13.2%) for primary repair.
- This analysis concluded that, Primary repair in the first year of life is the most prevalent strategy. Despite contemporary awareness of the late consequences of pulmonary insufficiency, ventriculotomy with transanular patch remains the most prevalent technique, both for primary repair and for repair following palliation.[7]

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Table 1. Discharge mortality stratified by type of procedure and age at surgery.

	0–30 Days	>30 Days–3 months	>3 Months–6 months	>6 Months–1 year	>1 Year–2 years	>2 Years–18 years	All ages
Tetralogy of Fallot palliation without previous cardiac surgery	11/179 = 6.1%	7/89 = 7.9%	2/16 = 12.5%	2/6 = 33%	0/1 = 0%	0/4 = 0%	22/295 = 7.5%
Tetralogy of Fallot palliation after previous cardiac surgery	0/4 = 0%	0/6 = 0%	0/1 = 0%	0/2 = 0%	0/1 = 0%	0/0 = 0%	0/14 = 0%
Tetralogy of Fallot repair without previous cardiac surgery	12/155 = 7.7%	8/492 = 1.6%	6/976 = 0.6%	4/614 = 0.6%	1/145 = 0.7%	2/154 = 1.3%	33/2536 = 1.3%
Tetralogy of Fallot repair after previous cardiac surgery	1/8 = 12.5%	0/10 = 0%	0/47 = 0%	1/111 = 0.9%	0/37 = 0%	0/4 = 0%	2/217 = 0.9%
All operations	24/346 = 6.9%	15/597 = 2.5%	8/1040 = 0.8%	7/733 = 1.0%	1/184 = 0.5%	2/162 = 1.2%	57/3062 = 1.9%

Table 1 documents discharge mortality stratified by type of procedure and age at surgery.[7] From these data, one can appreciate that in the current era in the United States of America, over 98% of patients who undergo repair of tetralogy of Fallot outside of the neonatal period will survive until hospital discharge. In all, 92.2% of neonates who undergo repair of tetralogy of Fallot will survive until hospital discharge.[7]

Management of late pulmonary insufficiency

Many patients will require late pulmonary valve replacement after surgery for tetralogy of Fallot. Indications for pulmonary valve replacement in this setting are evolving but currently include patients with moderate to severe pulmonary insufficiency and/or stenosis and any of the following problems:[8]

- Exertional symptoms of New York Heart Association class II or greater
- Decreased performance capacity on exercise testing
- Significant right ventricular dilation (>150 mm/m^2 by magnetic resonance imaging)
- Significant right ventricular dysfunction
- Significant ventricular arrhythmias
- QRS duration >180 ms

A variety of options exist for PVR including homografts, stented and stentless heterogafts, mechanical valves, and a polytetrafluoroethylene valve.

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