

Anaesthesia recommendations for patients suffering from

Tetralogy of Fallot

Disease name: Tetralogy of Fallot

ICD 10: XVII Q21.3

Synonyms: Fallot's tetralogy

Tetralogy of Fallot (TOF) is the commonest cyanotic congenital heart disease (CHD), with an incidence of 3 in 10,000 births, representing 10% of all CHDs. Whilst there is a spectrum of presentations and morphological variants, the classical description comprises a non-restrictive ventricular septal defect (VSD), an over-riding aorta, right ventricular outflow tract obstruction (RVOTO) with resultant right ventricular hypertrophy. Presentation is usually cyanosis and murmur in the neonatal period, although it can present later in milder forms.

Associations with chromosomal abnormalities are described, with microdeletion of 22q11.2 (cf. DiGeorge and velocardiofacial syndromes) being most frequent and trisomy 21, 13 and 18 also being overrepresented.

A hypercyanotic ('tet') spell is the feared complication of an unrepaired TOF, caused by excessive shunting, with potential lethal consequences. There is an excellent prognosis following surgical repair and most are asymptomatic. Despite this, there is excess mortality and morbidity due to chronic pulmonary regurgitation, recurrence of pulmonary stenosis, RV dysfunction, ventricular arrhythmias and sudden cardiac death.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong



Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

Complete repair of TOF normally takes place aged 6-18 months. Palliative procedures such as B-T (Blalock-Taussig) shunt may take place prior to this. Uncommonly, later repairs may occur in mild cases with delayed diagnosis.

Post-surgical correction, TOF patients may present to anaesthetists for procedures related to their disease, such as pulmonary valve (PV) replacement, Implantable Cardioverter Defibrillator (ICD) insertion and catheter interventions such as arrhythmia ablations. They may also be encountered on the labour ward as parturients, necessitating interventions for caesarean section or labour analgesia. With an expanding population of adult TOF patients, they are likely to present for non-cardiac surgery typical of the general population such as cholecystectomy and hysterectomy as well as for emergency surgery.

Type of anaesthesia

Pre-surgical repair:

The cardiac lesion in TOF predisposes to a right to left shunt and pulmonary hypoperfusion due to the presence of a RVOTO and unrestricted VSD. Concerns regarding the effect of a reduction in systemic vascular resistance (SVR) – that is, an increased right to left shunt with worsening hypoxaemia, lactatemia and cardiovascular collapse – has meant regional anaesthesia has classically been avoided, although it has been described.

General anaesthesia is the preferred mode of anaesthesia, and indeed a hypercyanotic spell resistant to medical management (manoeuvres to increase SVR, opiates and intravenous fluids), necessitates general anaesthesia, paralysis, intubation and positive pressure ventilation with control of SVR with phenylephrine or other vasopressor.

Inotropic support may be required, although in the context of a hypercyanotic spell adrenergic agents such as dobutamine and adrenaline are purported to worsen the causative infundibular spasm. Preparation for such an event may also involve beta blockers, which the patient may already be on. In severe cases, surgeon and perfusionist will need to be on standby.

Post-palliative procedure:

A palliative procedure such as B-T shunt is undertaken to maintain pulmonary perfusion in select patients. The aim of anaesthesia in such patients is to maintain patency of the shunt by ensuring good hydration and balancing and maintaining pulmonary (Qp) and systemic (Qs) perfusion. Worsening hypoxaemia in these patients is likely to be due to low SVR and low Qp, whilst 'normal' saturations indicate excess Qp and low Qs (systemic hypoperfusion and acidosis) and is also undesirable. Manipulation of SVR and PVR (pulmonary vascular resistant) is necessary in these scenarios.

Post-surgical repair:

Following correction of TOF, both regional and general anaesthetic techniques are well tolerated. However, residual deficits, long-term sequelae of pulmonary regurgitation (right ventricular dysfunction and arrhythmias) and aortic root dilatation and its consequences should be sought for and will influence the perioperative management.

Necessary additional diagnostic procedures (preoperative)

TOF patients will ideally be under cardiologists with special interest in congenital cardiac disease in a dedicated centre. Following complete surgical repair, there is annual follow-up eliciting long-term sequelae and its management.

Investigations include:

Bloods: cyanotic patients may be polycythaemic. Haematocrit > 0.65 may require venesection. A mild coagulopathy may also be present. Patients with B-T shunt (or equivalent) will be on Aspirin. Electrolyte imbalance and whole body magnesium depletion is possible with diuretic therapy.

ECG: Right bundle branch block is evident post-surgical repair. A QRS duration > 180ms is associated with malignant ventricular arrhythmias and RV dysfunction, seen in the older repaired patient.

Echo: Right ventricular function and dimensions, severity of pulmonary regurgitation. Left ventricular function (may be impaired due to incomplete myocardial protection in old surgery) and evidence of aortic regurgitation or root dilatation. Residual deficits such as VSD leak, residual RVOTO.

Cardiac MRI/CT: increasingly used to elicit cardiac morphology and function.

CPET: exercise intolerance can be quantified, and additionally provides prognostic information.

Holter monitor: Arrhythmias are common post repair, including interatrial re-entry tachycardias (Atrial flutter) and ventricular arrhythmias, often responsible for late sudden cardiac death.

Particular preparation for airway management

Whilst a specific association per se between difficult airway and TOF is not evident in the literature, its association with chromosomal abnormalities mean dysmorphic patients may be encountered. For example, Edward's (trisomy 18), Down's syndrome (trisomy 21) and DiGeorge syndrome (22q11 deletion) are known to pose a challenging airway on occasion. Consideration should be given for the presence of difficult airway equipment and strategy on an individual basis.

Tracheal anomalies are not uncommon (11%) and may require a range of smaller endotracheal tubes to be available. Perioperative complications resulting from these anomalies are likely.

Particular preparation for transfusion or administration of blood products

In cyanotic patients, polycythaemia and mild coagulopathy can be expected. Patients with B-T shunts or equivalent will be on Aspirin.

No specific requirements in non-cardiac surgery.

Particular preparation for anticoagulation

No specific requirements in non-cardiac surgery.

Patients with mechanical valve replacements (pulmonary or aortic) may be on anticoagulants.

Particular precautions for positioning, transport or mobilisation

No specific precautions.

In a suspected hypercyanotic spell in an unrepaired TOF, knee-to-chest position increases SVR (and reduces return of very deoxygenated blood from lower limbs) and thus reduces shunting and cyanosis. In the anaesthetised patient, this is achieved with vasopressors.

Probable interaction between anaesthetic agents and patient's long-term medication

Patients at high risk of hypercyanotic spells pre-repair may be on Propranolol. This blunts the effect of inotropic agents such as isoprenaline for 24h through competitive inhibition, although may not be clinically relevant.

Anaesthesiologic procedure

The pre-operative evaluation should assess the presence, frequency and severity of hypercyanotic spells. Heart failure is described although uncommon; features of cardiac insufficiency include tachypnoea, tachycardia, sweating, and cool peripheries – and in infants, poor feeding, failure to thrive and hepatomegaly.

Current or recent respiratory tract infections (implications for PVR) and functional status should be sought. Beta blockers are commenced in patients at risk of hypercyanotic spells and therefore its presence signals a higher risk. A history of associated syndromes and their intrinsic systemic effects may be elicited. Patients with long-standing disease may manifest complications associated with polycythaemia that include intracranial abscess, stroke and developmental delay.

A focussed cardiorespiratory examination is necessary; evaluation of oxygen saturations is pertinent, particularly where BT shunts are concerned. The morphology of the cardiac lesion and its effects should be appreciated, and investigations reviewed.

Avoidance of sympathetic stimulation with anxiolytic and sedative premedication is beneficial in those at risk of hypercyanotic spells. Examples of drugs that have been used successfully include midazolam (oral 0.5mg/kg; nasal 0.2mg/kg), chloral hydrate (oral 50mg/kg), pentobarbitone (rectal 2mg/kg), ketamine (intranasal 10mg/kg) and morphine (intramuscular 0.2mg/kg). Intramuscular morphine was associated with transient oxygen desaturation and

intranasal ketamine demonstrated improved separation scores and acceptance of intravenous cannulation (versus midazolam).

Intravenous induction with Ketamine has been advocated (vs inhalational induction) in the past, with the aim of preserving SVR and avoiding hypercyanotic attacks. However, it is generally accepted that the choice of drug or method of induction is less important than careful dose titration and duration over which it is given combined with an experienced anaesthetist. In our institution, inhalational induction is used frequently.

The cardiovascular effects of nitrous oxide include increased PVR and direct cardiodepressant effect (usually countered by its sympathomimetic effects). Whilst it has been used safely for induction and maintenance, where it reduces opiate requirements and haemodynamic response to sternotomy, it is perhaps best avoided in cases and where cardiac function is compromised significantly. Not used in our institution for cardiac surgery.

Opiates are well established for anaesthesia and analgesia. Non-steroidal anti-inflammatory drugs have been used; usual precautions apply e.g bleeding risk, renal dysfunction etc. Clearly, pre-corrected TOF patients on prostin infusions for a duct-dependent pulmonary circulation should avoid NSAIDs.

Antibiotics prophylaxis for infective endocarditis is advised for dental procedures in high risk cases. These include all pre-surgically corrected TOFs (including shunts/conduits). Post correction, those with previous infective endocarditis, residual deficits and within 6 months of prosthetic material use should be considered. Not recommended in respiratory tract, gastrointestinal, genitourinary, dermatological, or musculoskeletal procedures unless there is an established infection. [ESC guidelines]

Total intravenous anaesthesia (TIVA) techniques have been described. However, the cardiodepressant effects, reduction in SVR and propofol infusion syndrome (PRIS) phenomenon mean propofol should be used with care and in suitable patients.

Remifentanyl, ketamine and dexmedetomidine infusions have all been used for TIVA and adjuncts. Dexmedetomidine has the added advantage of suppressing post-correction junctional ectopic tachycardia.

Phosphodiesterase-3 inhibitors (milrinone, enoximone) are 'inodilators' with pulmonary vasodilator effects. They are useful perioperatively for TOF repair and in the presence of RV dysfunction, which may accompany pulmonary valve replacement.

Particular or additional monitoring

Pre-correction, post-palliative surgery and those with significant residual disease and/or long term effects should have invasive monitoring considered. Femoral arterial access is preferred due to the use of subclavian artery for shunts, which will also be clamped during its formation. Central venous lines enable administration of inotropes and guide fluid management. Transoesophageal echocardiography (TOE) and near-infrared spectroscopy (NIRS) may also be used, dictated by judgement and procedure.

Post-correction, invasive monitoring is instituted according to clinical judgement. For example, in one series of six elective caesarean sections, invasive arterial monitoring was used only in three cases (central venous monitoring in two). Patients who have had B-T shunt constructed may have compromised circulation and thus contralateral upper limb (or femoral) invasive monitoring is advised.

Possible complications

Air embolism: care with IV lines to avoid air bubbles and consider air filters (unrestricted or residual VSD)

Arrhythmias: junctional ectopic tachycardias, post correction. Later in disease: Interatrial re-entry tachycardia (flutter), Atrial Fibrillation, Ventricular Tachycardia and Ventricular Fibrillation possible.

Hypercyanotic attack: increased right to left shunting with pulmonary hypoperfusion due to infundibular spasm.

Congestive cardiac failure.

Right ventricular dysfunction.

Postoperative care

AHA/ACC recommended that patients with complex CHD are managed in a regional centre specializing in congenital cardiology, with experienced surgeons and cardiac anaesthetists. These patients should be managed postoperatively in an intensive care environment for continued monitoring (invasive, ECG telemetry etc.) with input from cardiologists experienced in CHD.

Post-correction, patients may be managed locally with guidance from regional centres, to which they will often be known. Postoperative care in a HDU/ITU environment is ideal.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the diseases, e.g.:

Hypercyanotic attack in an uncorrected TOF patient. These patients may be on propranolol as identified to be high risk. Aims of treatment are to reduce PVR and increase SVR and thus promote left to right shunting, and relieve infundibular spasm.

The patient will be deeply cyanosed, tachypnoeic and lethargic.

- Attempt knee-chest position
- IV fluid bolus
- Sedation (reduces catecholamines): IV/SC morphine, case reports of successful use of intranasal fentanyl and midazolam
- Paralyse and ventilate if unresponsive to these measures and commence phenylephrine.

Under anaesthesia, it will manifest as hypoxaemia, ischaemic ECG and hypotension.

Treatment:

- 100% O₂ (pulmonary vasodilator),
- *Light anaesthesia/sympathetic stimulation:*
 - Deepen anaesthesia,
 - IV fentanyl
 - IV propranolol/esmolol
- *Low SVR:*
 - IV Phenylephrine
 - IV fluid bolus

Ambulatory anaesthesia

Not suitable for ambulatory anaesthesia.

Obstetrical anaesthesia

Pregnancy in uncorrected TOF patients constitutes a significant risk of maternal and fetal morbidity and mortality. There are risks of right heart failure, arrhythmias and longer-term cardiovascular effects.

There is no consensus for the anaesthetic management of uncorrected TOFs and evidence is limited to case reports. There are advocates for low dose sequential CSE technique for brittle CHDs including TOFs, although both GA and conventional neuraxial techniques have been used. Whichever technique is utilised, a thorough understanding of the lesion and its cardiovascular effects must be appreciated. The usual anaesthetic aims remain: maintain preload and SVR, minimise PVR increases and avoid sympathetic stimulation (cf. pain and anxiety). Support from a regional centre is invaluable.

The risk in corrected TOF depends on residual disease burden and haemodynamics. Epidural anaesthesia is most often used in reported case series. There are increased perioperative complication rates of fetal death and maternal SVTs and cardiac failure. Despite these, favourable outcomes are reported.

The cardiovascular effects of uterotonics should be understood and its likely impact on TOF. Syntocinon causes hypotension, tachycardia and decreased cardiac output and temporary coronary insufficiency. Ergometrine given intravenously may cause vasoconstriction, hypertension and coronary vasospasm. Carboprost (PGF₂α) increases PVR, causes systemic hypotension with reflex tachycardia. As such, these should be used with caution balancing risk and benefit.

Although labour and delivery were previously considered high risk, antibiotics for infective endocarditis prophylaxis is now limited to patients likely to have bacteraemia at delivery.

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