Aktiv Druck & Verlag GmbH | ISSN 0170 - 5334 I 02330

www.ai-online.info

59. Jahrgang | Dezember 2018

A&I

ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

Offizielles Organ: Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V. (DGAI)
Berufsverband Deutscher Anästhesisten e.V. (BDA)
Deutsche Akademie für Anästhesiologische Fortbildung e.V. (DAAF)
Organ: Deutsche Interdisziplinäre Vereinigung für Intensiv- und Notfallmedizin e.V. (DIVI)

Tetralogy of Fallot

Thrombocytopenia-Absent Radius (TAR) syndrome

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 18 | 2018

OrphanAnesthesia –
ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –
a common project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

A survey of until now in A&I published guidelines can be found on:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

Projektleitung

Prof. Dr. Tino Münster, MHBA
Geschäftsführender Oberarzt
Facharzt für Anästhesie,
Spezielle Schmerztherapie,
Notfallmedizin
Anästhesiologische Klinik
Friedrich-Alexander-Universität
Erlangen-Nürnberg
Krankenhausstraße 12
91054 Erlangen, Deutschland
Tel.: 09131 8542441
Fax: 09131 8536147
E-Mail: muenster@kfa.imed.uni-erlangen.de

Deutsche Gesellschaft für Anästhesiologie & Intensivmedizin
www.dgai.de

A&I
ANÄSTHESIOLOGIE & INTENSIVMEDIZIN
www.ai-online.info

orphananesthesia

Anaesthesia recommendations for

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

▶ **Citation:** Haque M, Smith J: Tetralogy of Fallot. Anästh Intensivmed 2018;59:S654–S663.
DOI: 10.19224/ai2018.S654

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 an 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, whereas ‘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).

© Anästh Intensivmed 2018;59:S654-S663 Aktiv Druck & Verlag GmbH

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 in which 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.

Antibiotic 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.
Remifentanil, 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.

© Anästh Intensivmed 2018;59:S654-S663 Aktiv Druck & Verlag GmbH

Possible complications

Air embolism: Provide 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 are possible.

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

Congestive cardiac failure.

Right ventricular dysfunction.

Post-operative care

AHA/ACC recommended that patients with complex CHD are managed in a regional centre specialising 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% O_2 (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 foetal 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 the 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 foetal 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 (PGF2alpha) 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 are now limited to patients likely to have bacteraemia at delivery.

Literature and internet links

1. Adachi K, Ejima Y, Adachi O, Yamauchi M. Anesthetic management of pulmonary valve replacement for pulmonary regurgitation in six patients with surgically repaired tetralogy of Fallot. J Anesth 2014,28:928–931
2. Apitz C, Webb GD, Redington AN. Tetralogy of fallot. Lancet 2009;374:1462–1471
3. Arendt KW, Fernandes SM, Khairy P, Warnes CA, Rose CH, Landzberg MJ, et al: A case series of the anesthetic management of parturients with surgically repaired tetralogy of Fallot. Anesth Analg 2011;113:307–317
4. Juwakar C, Bharne S. Anaesthetic management of a parturient with uncorrected tetralogy of Fallot undergoing caesarean section. Anesth Essays Res 2012;6:244–246. DOI:10.4103/0259-1162.108358
5. Bailliard F, Anderson RH. Tetralogy of fallot. Orphanet J Rare Dis 2009;13;4:2
6. Barazzone C, Jaccard C, Berner M, Dayer P, Rouge JC, Oberhansli I, et al: Propranolol treatment in children with tetralogy of Fallot alters the response to isoprenaline after surgical repair. Br Heart J 1988;60:156–161
7. Bansal T, Saini S. Edward's syndrome: A rare cause of difficult intubation-utility of left molar approach. Egyp J Aneasth 2015;32:227–228
8. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al: ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31;23:2915–2957
9. Burt CC, Durbridge J. Management of cardiac disease in pregnancy. Continuing Education in Anaesthesia, Critical Care & Pain 2009;9;2:44–47
10. Byrd LA, Bruton-Maree N. Tetralogy of Fallot. AANA J 1989;57:169–176
11. Crean P, Koren G, Goresky G, Klein J, Macleod S. Fentanyl-oxygen versus fentanyl-N2O/oxygen anaesthesia in children undergoing cardiac surgery. Can Anaesth Soc J 1986;36–40
12. Dungan WT, Oztalay AY. Congestive Heart Failure in Tetralogy of Fallot: Report of Two Cases. Am J Dis Child 1964;107:398–403
13. Gelson E, Gatzoulis M, Steer PJ, Lupton M, Johnson M. Tetralogy of Fallot: maternal and neonatal outcomes. Br J Obstet Gynaecol 2008;115:398–402
14. Gharde P, Chauhan S, Kiran U. Evaluation of efficacy of intranasal midazolam, ketamine and their mixture as premedication and its relation with bispectral index in children with tetralogy of fallot undergoing intracardiac repair. Ann Card Anaesth 2006;9:25
15. Goyal R, Singh S, Bangi A, Singh SK. Case series: Dexmedetomidine and ketamine for anesthesia in patients with uncorrected congenital cyanotic heart disease presenting for non-cardiac surgery. J Anaesthesiol Clin Pharmacol 2013;29:543
16. Greeley WJ, Stanley TE, Ungerleider RM, Kisslo JA. Intraoperative Hypoxemic Spells in Tetralogy of Fallot. An Echocardiographic Analysis of Diagnosis and Treatment. Anesth Analg 1989;68:815–819
17. Hepburn L. Tetralogy of Fallot. Anaesthesia Tutorial Of The Week 2011;219
18. Juwarkar C, Bharne SS. Anesthetic management of a parturient with uncorrected tetralogy of Fallot for Cesarean section. Anesthesia 2012;6:244
19. Adinarayanan S, Parida S, Kavitha J, Balachander H. Spinal anesthetic for emergency cesarean section in a parturient with uncorrected tetralogy of Fallot, presenting with abruptio placentae and gestational hypertension. J Anaesthesiol Clin Pharmacol 2014;30:400–402. DOI: 10.4103/0970-9185.137276
20. Kadam SV, Tailor KB, Kulkarni S, Mohanty SR, Joshi PV, Rao SG. Effect of dexmeditomidine on postoperative junctional ectopic tachycardia after complete surgical repair of tetralogy of Fallot: A prospective randomized controlled study. Ann Card Anaesth 2015;18:323
21. Kaur H, Suri V, Aggarwal N, Chopra S, Vijayvergiya R, Talwar KK. Pregnancy in Patients with Tetralogy of Fallot Outcome and Management. World J Pediatr Congenit Heart Surg 2010;1:170–174
22. Kazim R, Quaegebeur JM, Sun LS. The association of tracheal anomalies and tetralogy of Fallot. J Cariothorac Vasc Anesth 1996;10:589–592
23. Levine ME, Hartley EJ, Macpherson BA, Burrows FA, Lerman J. Oral midazolam premedication for children with congenital cyanotic heart disease under-going cardiac surgery: a comparative study. Can J Anaesth 1993;40:934–938

© Anästh Intensivmed 2018;59:S654-S663 Aktiv Druck & Verlag GmbH

24. Lovell AT. Anaesthetic implications of grown-up congenital heart disease. Br J Anaesth 2004;93:129–139
25. Montero JV, Nieto EM, Vallejo IR, Montero SV. Intranasal Midazolam for the Emergency Management of Hypercyanotic Spells in Tetralogy of Fallot. Pediatr Emerg Care 2015;3:269–271
26. Spielman FJ, Herbert WN. Maternal cardiovascular effects of drugs that alter uterine activity. Obstet Gynecol Surv 1998;43:516–522
27. Thomas JS, Koh SH, Cooper GM. Haemodynamic effects of oxytocin given as iv bolus or infusion on women undergoing Caesarean section. Br J Anaesth 2007;98:116–119
28. Tsze DS, Vitberg YM, Berezow J, Starc TJ, Dayan PS. Treatment of Tetralogy of Fallot Hypoxic Spell With Intranasal Fentanyl. Pediatrics 2014;134:266–269
29. Villafañe J, Feinstein JA, Jenkins KJ, Vincent RN, Walsh EP, Dubin AM, Dearani J. Hot topics in tetralogy of Fallot. J Am Coll Cardiol 2013;62:2155–2166
30. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al: ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease) Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeon. J Am Coll Cardiol 2008;52(23):143–263
31. White MC, Peyton JM. Anaesthetic management of children with congenital heart disease for non-cardiac surgery. Continuing Education in Anaesthesia Critical Care and Pain 2012;12: 17–22.

Last date of modification: July 2016

This guideline has been prepared by:

Authors
Mohammed Haque, Anaesthetic registrar, Great Ormond Street Hospital, London, United Kingdom
ebadulhaque@doctors.org.uk

Jonathan Smith, Consultant paediatric anaesthetist, Great Ormond Street Hospital, London, United Kingdom

Peer revision 1
Ozkan Onal, Department of Anesthesia and Intensive Care, Selcuk University Medical Faculty, Konya, Turkey
drozkanonal@selcuk.edu.tr

Peer revision 2
Adeola Animasahun, Department of Paediatrics and Child Health, Lagos State University College of Medicine, Ikeja, Lagos, Nigeria
deoladebo@yahoo.com

© Anästh Intensivmed 2018;59:S654-S663 Aktiv Druck & Verlag GmbH

Herausgeber

DGAI
Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V.
Präsident: Prof. Dr. B. Zwißler, München

BDA
Berufsverband Deutscher Anästhesisten e.V.
Präsident: Prof. Dr. G. Geldner, Ludwigsburg

DAAF
Deutsche Akademie für Anästhesiologische Fortbildung e.V.
Präsident: Prof. Dr. F. Wappler, Köln

Korrespondenzadresse: Roritzerstraße 27 | 90419 Nürnberg | Deutschland
Tel.: 0911 9337812 | Fax: 0911 3938195
E-Mail: anaesth.intensivmed@dgai-ev.de

Verlag & Druckerei

Aktiv Druck & Verlag GmbH
An der Lohwiese 36 |
97500 Ebelsbach | Deutschland
www.aktiv-druck.de

Geschäftsführung
Wolfgang Schröder | Jan Schröder | Nadja Schwarz
Tel.: 09522 943560 | Fax: 09522 943567
E-Mail: info@aktiv-druck.de

Anzeigen | Vertrieb
Pia Engelhardt
Tel.: 09522 943570 | Fax: 09522 943577
E-Mail: anzeigen@aktiv-druck.de

Verlagsrepräsentanz
Jürgen Distler
Roritzerstraße 27, 90419 Nürnberg
Tel.: 0171 9432534 | Fax: 0911 3938195
E-Mail: jdistler@bda-ev.de

Herstellung | Gestaltung
Manfred Wuttke | Stefanie Triebert
Tel.: 09522 943571 | Fax: 09522 943577
E-Mail: ai@aktiv-druck.de

Titelbild
Dipl.-Designerin Monique Minde, Nürnberg

Erscheinungsweise 2018
Der 59. Jahrgang erscheint jeweils zum Monatsanfang, Heft 7/8 als Doppelausgabe.

Bezugspreise (inkl. Versandkosten):

- **Einzelhefte** 30,- €
- **Jahresabonnement:**
  Europa (ohne Schweiz) (inkl. 7 % MwSt.) 258,- €
  Schweiz 266,- €
  Rest der Welt 241,- €

Mitarbeiter aus Pflege, Labor, Studenten und Auszubildende (bei Vorlage eines entsprechenden Nachweises)
Europa (ohne Schweiz) (inkl. 7 % MwSt.) 94,- €
Schweiz 90,- €
Rest der Welt 94,- €

Für Mitglieder der DGAI und/oder des BDA ist der Bezug der Zeitschrift im Mitgliedsbeitrag enthalten.

Allgemeine Geschäfts- und Lieferbedingungen
Die allgemeinen Geschäfts- und Lieferbedingungen entnehmen Sie bitte dem Impressum auf www.ai-online.info

Indexed in **Current Contents®/Clinical Medicine, EMBASE/Excerpta Medica; Medical Documentation Service; Research Alert; Sci Search; SUBIS Current Awareness in Biomedicine; VINITI: Russian Academy of Science.**

Online-Ausgabe der A&I ab April 2017 open access: **www.ai-online.info**

CONTACT US

Please do not hesitate to contact us. We will be glad to answer and provide further information to you at any time.

.....
Name

.....
First Name

.....
Department / Hospital

.....
Place

.....
Telephone

.....
E-Mail

.....
Date / Signature

Please contact me for further information ○

I would like to participate in the project ○

ADDRESS

German Society of Anaesthesiology and
Intensive Care Medicine
Ursula Homberg
Roritzerstrasse 27 | 90419 Nuremberg | Germany
Tel.: +49-911-9337828 | Fax: +49-911-3938195
Email: uhomberg@orphananesthesia.eu