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Dubowitz syndrome

Eisenmenger syndrome

orphananesthesia

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

SUPPLEMENT NR. 8 | 2021

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
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OrphanAnesthesia –

a 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.

Find a survey of the recommendations published until now on:

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Anaesthesia recommendations for

Eisenmenger syndrome

Disease name: Eisenmenger syndrome

ICD 10: Q21.8

Synonyms: Eisenmenger disease; Eisenmenger complex

Disease summary: Eisenmenger syndrome develops in patients with left-to-right shunts that result in right heart volume overload. The shunt is most commonly due to atrial septal or ventricular septal defects, although any intracardiac defect that results in left-to-right shunting of blood can result in volume and pressure overload. Increased pulmonary vascular resistance is a consequence of this volume overload, which results in right ventricular enlargement, pulmonary hypertension, and reversal of the left-to-right shunt into a bidirectional or right-to-left fixed shunt. Patients are clinically cyanotic, frequently have dyspnoea on exertion, decreased exercise tolerance, signs and symptoms of congestive heart failure including signs of right ventricular overload and failure, peripheral oedema, syncope, and may have alterations in end-organ function (hepatic congestion, abnormal bleeding, cerebral vascular accidents, renal failure, etc.). Patients may be normal in appearance or may be syndromatic. Pregnancy carries a particularly high peripartum risk for premature labour as well as mortality regardless of delivery method and is discouraged. Patients may present in the 3rd and 4th decades of life, although smaller shunts may not be problematic until later in life. Patients are often managed with pulmonary vasodilators to reduce the pulmonary vascular resistance, improve quality of life and exercise tolerance, as well as treat heart failure, if present.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

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Typical surgery

Patients may present for any and all elective and emergent surgeries. Some of the more common indications for elective surgery include: dental procedures; gynaecologic procedures including examinations under anaesthesia for routine preventive care and tubal ligations; left and right heart catheterisation; transoesophageal echocardiogram; appendectomy; cholecystectomy.

Type of anaesthesia

Monitored anaesthetic care (MAC); general anaesthesia; regional anaesthesia.

Necessary additional pre-operative testing (beside standard care)

Recent echocardiogram.

Type of congenital heart defect and shunt directionality (fixed or bidirectional; anatomical considerations; additional cardiac defects).

Electrocardiogram (right ventricular hypertrophy with conduction delay or various types of heart block is frequently seen).

Presence of intrinsic defibrillators (ICD) or pacemakers and indication for placement.

If an ICD or pacemaker is present, a thorough evaluation of the device, indication, settings, and recent defibrillation(s) for ICDs should be known. Interrogation of the device by trained personnel (device representative, cardiologist, etc.) is required to assess these devices and adjust parameters depending on indication, surgical location, and pacing requirements. ICDs should be deactivated and pacemakers may need to be adjusted for rate responsiveness or need for asynchronous pacing (VOO/DOO).

Baseline oxygen saturation with and without supplemental oxygen.

Right heart catheterisation to assess for pulmonary vasoreactivity.

Despite Eisenmenger syndrome being associated with “fixed” reactivity, some patients’ pulmonary vascular resistance will decrease with selective pulmonary vasodilators and can be demonstrated on right heart catheterisation. This is not mandatory for noncardiac surgery, however, and should not result in delay or cancellation of an operative procedure.

Particular preparation for airway management

Airway preparation should be based on each patient and his or her underlying disease process including assessment of any possible congenital malformation. Some patients will have trisomy 21 and may present with glossal enlargement, reduced head and neck mobility, concern for atlanto-occipital instability, and reduced oral opening. Patients with neurologic delay or disabilities may not cooperate during awake intubation, so judicious premedication to obtain a cooperative, but spontaneously breathing patient will be mandatory if there is concern for difficulty with standard airway management. Due to these concerns, many

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uncooperative patients will tolerate inhalational induction while maintaining spontaneous respirations. For patients in an obstetric environment, a difficult airway kit should be immediately available due to potential for further airway impairment related to pregnancy. Care should be taken to avoid precipitators of pulmonary vascular resistance, which may worsen shunt fraction.

Factors that increase pulmonary vascular resistance:

1) increased catecholamines (elevated sympathetic surge, light anaesthesia, etc.)
2) hypoxaemia via hypoxic pulmonary vasoconstriction
3) hypercarbia
4) lung distention (optimal PVR at functional residual capacity)
5) acidosis
6) hypothermia

Particular preparation for transfusion or administration of blood products

Type and crossmatch is not mandatory, but patients frequently have an extensive surgical and transfusion history. An antibody screen prior to surgery with appropriate reservation of blood products is recommended for surgical procedures that are moderate or high risk. Coagulation testing may be abnormal as hepatic function is often altered in cyanotic diseases. Additionally, patients are often on anticoagulants to prevent microvascular thrombosis.

Particular preparation for anticoagulation

None other than standard.

Particular precautions for positioning, transportation and mobilisation

None beyond what is specific to patient or disease.

Interactions of chronic disease and anaesthesia medications

None reported.

Anaesthetic procedure

The induction and maintenance of anaesthesia in patients presenting for noncardiac surgery with Eisenmenger syndrome should focus on maintaining pre-operative baseline haemodynamics, avoiding hypotension, which can worsen the right-to-left shunt, and providing adequate gas exchange via the lungs to reduce shunting, acidosis, and cardiac

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decompensation. Pharmacological management of patients with Eisenmenger physiology aims to attenuate the elevated pulmonary vascular resistance. Current medical therapies include oral phosphodiesterase-5 inhibitors (sildenafil, etc.), prostanoids (epoprostenol, treprostinil, iloprost), and endothelin-1 receptor antagonists (bosentan). These medications can be delivered via the IV, subcutaneous, inhalational, or oral route and should be continued peri-operatively [1,2]. Intra-operative management with inhaled pulmonary vasodilators (nitric oxide, epoprostenol), along with TEE, have been successfully used in patients undergoing urgent surgery and are acceptable therapies for elective as well as urgent/emergent operations to potentially reduce pulmonary vascular resistance [3]. The actual intra-operative effects on PVR with use of these agents are not known, however, and are only hypothesised. Nitric oxide remains expensive with a difficult delivery system and may not be as beneficial, particularly as reductions in pulmonary vascular resistance may not occur in patients with fixed physiology [4].

Induction and maintenance of general anaesthesia should be limited to anaesthetic induction agents that result in the least degree of haemodynamic compromise. Frequently used drugs include ketamine and etomidate with supplemental opioids to attenuate the sympathetic surge during stimulation and intubation. Induction of general anaesthesia should be done rapidly to avoid periods of hypoventilation, but should not allow significant hypotension to occur. A bolus of a vasopressor at the time of induction was shown to be beneficial in reducing hypotension in patients receiving general anaesthesia and should be considered [5]. An infusion of vasopressor may be ideal for maintenance of SVR during induction as well as throughout the anaesthetic process. To further optimise the goal of maintenance of systemic vascular resistance, a pre-induction arterial catheter placed under local anaesthesia is beneficial and recommended.

Maintenance of general anaesthesia may be performed with either volatile agents or total intravenous anaesthesia (TIVA). Volatile agents are known to decrease systemic vascular resistance with less effect on the pulmonary system. Of potential benefit is the decreased response to hypoxaemia via the hypoxic pulmonary vasoconstrictive response, which is blunted by volatile anaesthesia. Sevoflurane may be ideal due to decreased pungency allowing for inhalational induction and patient tolerance. Furthermore, sevoflurane has known beneficial effects on pulmonary bronchial reactivity, which can exacerbate pulmonary resistance. N2O has little effect on pulmonary blood flow, but does cause pulmonary vasoconstriction, which increases PVR. The effects of TIVA are less known, though the effects on hypoxic pulmonary vasoconstriction are less than volatile agents. The decrease in SVR during TIVA may be more pronounced than with volatile agents and drug concentration should be titrated with use of the bispectral index (BIS). For either method of maintenance, opioids need to be judiciously given to reduce marked effects on respiration (hypoventilation, hypercarbia, decreased ventilatory responsiveness to hypoxia, etc.). Pain needs to be managed to decrease the catecholamine surge present in patients with poorly controlled pain and multimodal analgesia with use of non-steroidal anti-inflammatory drugs, regional anaesthesia, and other agents may be considered to help reduce opioid requirements.

General anaesthesia with laparoscopic surgery poses a particular increased risk in Eisenmenger syndrome due to insufflation with carbon dioxide (CO2) under pressure. This may result in hypercapnoea that is difficult to manage, resulting in respiratory acidosis, potential worsening of the right-to-left shunt with subsequent hypotension, arterial deoxygenation, and ultimately cardiac decompensation. The anaesthesiologist is advised to maintain normocapnia, which may require hyperventilation prior to insufflation to reduce the PaCO2 sufficiently prior to CO2 insufflation. Furthermore, the surgeon should be instructed to insufflate the peritoneal cavity with as little pressure as possible to safely perform the surgery in order to facilitate adequate venous return. Discussion with the surgeon on need to convert to open should be done and criteria agreed upon prior to surgical start. If arterial catheters are present, arterial blood gases should be regularly reviewed to help guide

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ventilator adjustments as well as the decision to convert to open surgery. Intra-abdominal pressure increases can displace the diaphragm and result in malposition of the endotracheal tube into a mainstem bronchus. Peak airway pressures may rapidly rise from the elevated abdominal pressure or endotracheal tube malposition, which may reduce effective venous return. Trendelenburg positioning can further exacerbate these intra-operative events, resulting in acute decompensation. Open surgical procedures may be required if the patient does not tolerate any portion of the laparoscopic procedure and the surgical team should be ready to rapidly reduce the intra-abdominal pressure if haemodynamic compensation occurs.

Monitored anaesthetic care (MAC) is often considered for care of patients presenting for noncardiac surgery with Eisenmenger syndrome due to the perceived safety over general anaesthesia. However, oxygen desaturations may occur more commonly than in general anaesthesia along with hypercarbia due to obstruction, hypoventilation, and lack of adequate airway support. A recent study demonstrated that, in noncardiac surgical patients with Eisenmenger syndrome, 67% of recorded intra-operative oxygen desaturations occurred during MAC versus general anaesthesia [5]. Furthermore, patients with disabilities may not tolerate sedation well with potential agitation and combativeness, resulting in risk to the patient and surgical staff as well as sympathetic discharge and potential worsening of pulmonary vascular resistance and right-to-left shunting. Strict assessment and evaluation of the patient's sedation requirements and respiratory status should be performed, with medications and airway supplies immediately available and ready if a need to convert from MAC to general anaesthesia is encountered. Additionally, consideration for pre-induction arterial access with frequent arterial blood gases may be warranted to assess for ineffective respiratory efforts.

Patients may be particularly non-cooperative due to other mental and physical handicaps. Consideration for inhalational anaesthetic induction in these uncooperative and combative patients may seem ideal with intravascular cannulation obtained once the patient is asleep. While this helps reduce anxiety and catecholamine excess, patients may decompensate at any time or prove to be particularly challenging to appropriately ventilate through bag-mask ventilations. Peripheral IV access obtained prior to induction is preferable in patients presenting for any surgical procedure to facilitate rapid induction and treatment of haemodynamic decompensation. However, some patients are not cooperative enough to allow IV access and require inhalational induction. For these patients it is advisable to have a colleague present to place an IV once the patient has been anaesthetised. Pre-operative oral medications or intramuscular medications to facilitate a more docile and cooperative patient, while monitoring respiratory status, are advocated over inhalational induction without vascular access.

Regional anaesthesia has been successfully used in noncardiac surgery [6], in addition to parturients [7], and provides an attractive alternative to general anaesthesia. Selective nerve blocks can be used with judicious sedation to provide surgical level anaesthesia. Epidural anaesthesia is preferred over subarachnoid block due to the ability to slowly titrate the anaesthetic level while monitoring haemodynamics closely. Spinal anaesthesia is contraindicated in this patient population due to the rapid and often unpredictable level of anaesthesia. Particularly concerning is the potential for significant hypotension and cardiac decompensation which can dramatically result in worsening of the shunt fraction. Epidural anaesthesia should be slowly increased while avoiding preparations that contain epinephrine due to concern for significant effect on the pulmonary vascular resistance if an intravascular injection were encountered.

In any anaesthetic management, acute haemodynamic decompensation may manifest with oxygen desaturation and potential refractory hypotension [8,9]. Hypotension is often more likely the initial derangement, due to exacerbation of the right to left shunt. Oxygen desaturation frequently occurs after hypotension is observed as the shunt worsens. Use of

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phenylephrine is often the drug of choice for many anaesthetists to treat or prevent hypotension, though the effect on the pulmonary vasculature with a pure alpha-agonist may warrant consideration of mixed agents, like ephedrine or norepinephrine. Vasopressin is another potential effective vasopressor due to reduced effect on the pulmonary vascular bed while maintaining systemic vascular resistance [10]. Further treatment beyond vasopressor delivery includes correcting hypercarbia, hypoxia, acidosis, and hypothermia, as these are all known triggers for worsening of pulmonary hypertension [11,12]. Furthermore, evaluation for inadequate anaesthesia or analgesia should be performed as excessive sympathetic response can cause an increase in PVR.

Particular or additional monitoring

Invasive monitoring should be performed based on each patient and the surgical procedure. Invasive blood pressure monitoring with arterial catheters are beneficial to maintain strict blood pressure control, obtain frequent arterial blood gases intra-operatively as well as post-operatively, and are highly recommended. Arterial catheters should be placed prior to induction with local anaesthesia to facilitate optimal anaesthetic induction and maintenance of systemic blood pressure at baseline. Central venous catheters are not recommended for routine procedures unless peripheral intravenous access is difficult or unacceptable for patient care. Ultrasound guidance should be utilised to confirm anatomy and assess for venous thrombosis when central venous cannulation is performed. Further consideration of cardiac output monitoring with oesophageal Doppler or pulse-pressure based monitors (i.e. FloTrac, PhysioFlow, etc.) can be beneficial in evaluating volume status, cardiac output, and systemic vascular resistance.

Pulmonary arterial catheters are not advised and are problematic. There is significant risk with placement of a pulmonary arterial catheter in patients with right-to-left shunts and includes pulmonary artery rupture, paradoxical left sided placement with increased thrombotic/stroke risk, pathologic ventricular arrhythmia, and a significantly high failure rate without expert guidance and fluoroscopy due to abnormal anatomy with intracardiac defect.

Possible complications

Patients with Eisenmenger syndrome are at risk for venous thrombosis and pulmonary embolism due to hyperviscosity of blood.

Increased risk for respiratory compromise during immediate and delayed post-operative course, including dramatic worsening of respiratory status with scheduled pain medications.

Difficulty with vascular access including difficult central and arterial cannulation from repeated catheter placement. Ultrasound may be required to evaluate potential IV insertion and cannulation sites.

Medication mismanagement resulting in worsening of heart function and pulmonary hypertension/shunt.

Simplify the anaesthetic management plan as much as possible to help to reduce confusion between patient physiological derangement and anaesthetic induction/maintenance of complication.

Difficult intubation is possible related to additional comorbid conditions.

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Pulmonary arterial rupture during pulmonary artery catheter placement, including balloon inflation. This may occur during the intra-operative as well as the post-operative course.

Difficulty obtaining matched blood due to preformed antibodies. Particularly concerning in operations where haemorrhage may be encountered.

Poor patient cooperation resulting in inability to obtain peripheral IV access.

Post-operative care

Post-operative care should be performed in areas equipped for monitoring ECG, intermittent as well as continuous blood pressure (arterial pressure transduction capabilities), oxygen saturations, and temperature [13]. Post-operative mechanical ventilation may be required and patients should be cared for in an area well equipped to assess and manage ventilated patients, including arterial blood gas analysis. The post-anaesthesia care unit (PACU) may be adequate for patients following noncardiac surgical procedures and can facilitate same-day procedures. The intensive care unit (ICU) may be chosen for patients with large operative procedures, high fluid shift expectations, need for post-operative mechanical ventilation, and/or PACU inability to effectively manage any aspect of the patient's care. Patients housed in non-ICU hospital rooms should be admitted to floors where oxygen saturations and ECGs can be continuously monitored. Post-operative care should follow the same considerations and goals as those made pre- and intra-operatively. Post-operative hypoventilation and hypoxaemia, arrhythmias and poorly controlled pain may result in exacerbation of the patient's shunt, which could result in cardiac decompensation. Particular consideration regarding multimodal analgesia should be given to reduce opioid requirements. Patient controlled analgesia should be managed in intensive care settings due to decreased ventilatory response to hypoxia and hypercarbia. Parturients should be observed in an intensive care setting due to the acute physiologic changes immediately postpartum [14].

Disease-related acute problems and effect on anaesthesia and recovery

Eisenmenger syndrome may result in acute, life-threatening haemodynamic and respiratory decompensation. Oxygen desaturation beyond the baseline value, with or without hypotension, is frequently encountered. This may reflect a poor respiratory effort or worsening of the right-to-left shunt. Additional oxygen and manoeuvres to improve respiration will not improve the oxygen saturations if a shunt is the causative factor.

Hypotension may be the initial derangement of vital signs in a patient with worsening pulmonary vascular resistance. This may reflect the increase in right-to-left shunting or cardiac decompensation and should be treated aggressively.

Cardiac arrhythmias, including unstable ventricular arrhythmias, may be encountered. These may indicate worsening cardiac function, oxygenation, or reduced effective coronary perfusion, particularly in patients with thickened right ventricles.

Ambulatory anaesthesia

Patients with Eisenmenger syndrome should not be scheduled for ambulatory anaesthesia in an outpatient facility. Due to the nature of their disease, their potential need for invasive

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monitoring as well as cardiac decompensation, all surgical procedures should be performed in a hospital setting with the ability to manage the adult patient with congenital heart disease.

Obstetrical anaesthesia

Patients with Eisenmenger syndrome have an increased risk for peripartum mortality as well as premature labour. Of all pregnancies, 20-30% will result in spontaneous abortion, whereas premature delivery prevails in almost one-half of the remaining cases [14,15]. The changes in cardiac output, haemodynamics, as well as coagulation status increase the risk for sudden cardiac death, stroke, and postpartum cardiomyopathy leading to death in 30-45% of patients [14]. Patients are at increased risk for peripartum deep venous thrombosis formation as well as pulmonary embolism due to polycythaemia, venous congestion and procoagulant state of pregnancy.

Epidural anaesthesia has been shown to be effective in patients with Eisenmenger syndrome, but it requires judicious administration of anaesthetics to reduce the risk of hypotension with concomitant exacerbation of the right-to-left shunt. Arterial catheterisation is advisable in patients who receive an epidural to monitor haemodynamics closely while titrating the anaesthetic level. Central venous catheters may be necessary if peripheral intravenous access is not sufficient. Pulmonary arterial catheters are not recommended.

Patients should have an active type and screen due to risk for haemorrhage and need for transfusion to maintain volume status as well as systemic vascular resistance. Due to polycythaemia from chronic cyanosis, patients may tolerate more blood loss than healthy parturients before transfusion is required.

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