

## Anaesthesia recommendations for patients suffering from

### **Eisenmenger's Syndrome**

**Disease name:** Eisenmenger's syndrome

**ICD 10:** Q21.8

**Synonyms:** Eisenmenger's disease; Eisenmenger's complex

**Disease summary:**

Eisenmenger's 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, though 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 dyspnea on exertion, decreased exercise tolerance, signs and symptoms of congestive heart failure including signs of right ventricular overload and failure, peripheral edema, 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 syndromal. Pregnancy carries a particularly high peripartum risk for premature labor as well as mortality regardless of delivery method and is discouraged. Patients may present in the 3<sup>rd</sup> and 4<sup>th</sup> decades of life, though 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.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)

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## Typical surgery

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

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## Type of anaesthesia

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

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## Necessary additional diagnostic procedures (preoperative)

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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 catheterization to assess for pulmonary vasoreactivity

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

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## Particular preparation for airway management

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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 is mandatory if there is concern for difficulty with standard airway management. Due to these concerns, many uncooperative

patients will tolerate inhalational induction while maintaining spontaneous respirations. For patients in the 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 anesthesia, etc.)
- 2) hypoxemia via hypoxic pulmonary vasoconstriction
- 3) hypercarbia
- 4) lung distention (optimal PVR at functional residual capacity)
- 5) acidosis
- 6) hypothermia
- 7) increased blood viscosity

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**Particular preparation for transfusion or administration of blood products**

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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.

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**Particular preparation for anticoagulation**

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None other than standard.

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**Particular precautions for positioning, transport or mobilisation**

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None beyond patient/ disease specific.

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

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None reported.

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**Anaesthesiologic procedure**

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The induction and maintenance of anaesthesia in patients presenting for noncardiac surgery with Eisenmenger's syndrome should focus on maintaining preoperative baseline hemodynamics, avoiding hypotension, which can worsen the right-to-left shunt, and providing adequate gas exchange via the lungs to reduce shunting, acidosis, and cardiac decompensation. Pharmacologic management of patients with Eisenmenger's 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 perioperatively [1,2]. Intraoperative 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 the pulmonary vascular resistance [3]. The actual intraoperative effects on PVR are not known, however, with use of these agents and are only hypothesized. 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 hemodynamic compromise. Frequently used drugs include ketamine and etomidate with supplemental opioids to attenuate the sympathetic surge during stimulation and intubation. Induction of general anesthesia should be done rapidly to avoid periods of hypoventilation, but should not allow significant hypotension to occur. A bolus of a vasopressor at time of induction was shown to be beneficial in reducing hypotension in patients receiving general anesthesia and should be considered<sup>5</sup>. An infusion of vasopressor may be ideal for maintenance of SVR during induction as well as throughout the anesthetic. To further optimize the goal of maintenance of systemic vascular resistance, a preinduction 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 hypoxemia 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. N<sub>2</sub>O 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 Bispectral Index (BIS). For either method of maintenance, opioids need to be judiciously given to reduce marked effects on respiration (hypoventilation, hypercapnia, 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 antiinflammatory 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's syndrome due to insufflation with carbon dioxide (CO<sub>2</sub>) under pressure. This may result in hypercapnea 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 PaCO<sub>2</sub> sufficiently prior to CO<sub>2</sub> 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 performed and criteria agreed upon prior to surgical start. If arterial catheters are present, arterial blood gases should be regularly reviewed to help guide ventilator adjustments as well as the decision to convert to open. 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 intraoperative 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 hemodynamic compensation occurs.

Monitored anaesthetic care (MAC) is often considered for care of patients presenting for noncardiac surgery with Eisenmenger's 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's syndrome, 67% of recorded intraoperative 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 requirement 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 preinduction 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 hemodynamic 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 anesthetized. Preoperative 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 hemodynamics 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 hemodynamic 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 phenylephrine is often the drug of choice for many anesthetists 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 anesthesia or analgesia should be performed as excessive sympathetic response can cause an increase in PVR.

### **Particular or additional monitoring**

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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 intraoperatively as well as postoperatively, and are highly recommended. Arterial catheters should be placed prior to induction with local anesthesia 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 utilized to confirm anatomy and assess for venous thrombosis when central venous cannulation is performed. Further consideration of cardiac output monitoring with esophageal doppler or pulse-pressure based monitors (ie 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 significantly high failure rate without expert guidance and fluoroscopy due to abnormal anatomy with intracardiac defect.

### **Possible complications**

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

Increased risk for respiratory compromise during immediate and delayed postoperative 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 anaesthetic management plan as much as possible to help reduce confusion between patient physiologic derangement or anesthetic induction/ maintenance of complication.

Difficult intubation is possible related to additional comorbid conditions.

Pulmonary arterial rupture during pulmonary artery catheter placement, including balloon inflation. May occur during the intraoperative as well as postoperative 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.

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### **Postoperative care**

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Postoperative care should be performed in areas equipped for monitoring EKG, intermittent as well as continuous blood pressure (arterial pressure transduction capabilities), oxygen saturations, and temperature [13]. Postoperative 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-anesthesia 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 postoperative 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 EKGs can be continuously monitored. Postoperative care should follow the same considerations and goals as those made pre and intraoperatively. Postoperative hypoventilation and hypoxemia, 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 performed 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].

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### **Information about emergency-like situations / Differential diagnostics**

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Eisenmenger's 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 poor respiratory effort or worsening of the right-to-left shunt. Additional oxygen and maneuvers 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 of the 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.

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### **Ambulatory anaesthesia**

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

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Patients with Eisenmenger's syndrome have an increased risk for peripartum mortality as well as premature labor. Of all pregnancies, 20-30% will result in spontaneous abortion with the remaining having a rate of almost half being premature [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 polycythemia, venous congestion, and procoagulant state of pregnancy.

Epidural anaesthesia has been shown to be effective in patients with Eisenmenger's syndrome, but requires judicious administration of anesthetic to reduce the risk of hypotension with concomitant exacerbation of the right-to-left shunt. Arterial catheterization is advisable in patients who receive an epidural to monitor hemodynamics closely while titrating anesthetic 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 polycythemia from chronic cyanosis, patients may tolerate more blood loss than healthy parturients before transfusion is required.

## Literature and internet links

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