orphananesthesia

Anaesthesia recommendations for patients suffering from

Glucose-6-phosphate dehydrogenase deficiency

Disease name: Glucose-6-phosphate dehydrogenase deficiency

ICD 10: D55.0

Synonyms: Favism, G6PD deficiency, glucosephosphate dehydrogenase deficiency

Glucose-6-phosphate dehydrogenase deficiency (G6PD deficiency) is an enzymopathy of red blood cells in humans [18]. It is an X-linked, hereditary genetic defect, prevalent in up to 400 million people worldwide mainly in about 10% of African-Americans as well as to a lower frequency in the Mediterranean people [15,18]. It is known that G6PD-deficient cells protect against the malaria parasite Plasmodium falciparum in women by means of slowest parasite growing in these cells or earlier phagocytosis by macrophages [9,10,41,46]. G6PD is an enzyme necessary for the production of antioxidants, which protect red blood cells from oxidative stressors [18]. In case of G6PD deficiency, red blood cells can be damaged by oxidative stresses from certain drugs, metabolic conditions (diabetic ketoacidosis, metabolic acidosis), infections [5,28,44], hypothermia, lawsone (Henna), ingestion of fava beans or stress related to surgical interventions, resulting in haemolysis [17,18].

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Disease summary

The severity of disease is determined by the magnitude of enzyme deficiency: Class I variants have severe enzyme deficiency with chronic non-spherocytic haemolytic anaemia (<10% residual enzyme activity), class II variants also have severe enzyme deficiency (<10% residual enzyme activity) but with intermittent acute haemolysis, class III variants have moderate enzyme deficiency (10-60% residual enzyme activity) with intermittent acute haemolysis. Class IV and V variants are of no clinical significance - class IV has no enzyme deficiency and class V has increased enzyme activity [18]. However, severity is more pronounced in G6PD deficient white patients (acute renal failure) than in the black population (selflimitation) [10,11]. Haemolysis is influenced by the type of mutation causing disease, genetic make-up and gender of the individual, age of erythrocytes, the type and dose of offending drug [44] and the number of other present risk factors, for example infections [39].

The main anaesthetic concern in the treatment of patients with G6PD deficiency is the choice of drugs using for anaesthetic management, and postoperative pain therapy since exposure to oxidative drugs can lead to haemolytic crisis in these patients.

Pathogenesis

The G6PD is a key enzyme in glucose metabolism [6,10,15]. G6PD converts glucose-6-phosphate to 6-phosphogluconolactone combined with the reduction from NADP (nicotinamide-adenine dinucleotide phosphate) to NADPH [6,10,15]. In the red blood cell this pathway is the only source of NADPH. NADPH is required for reduction of glutathione [6,10,15]. The reduced form of glutathione is essential for the protection against oxidation of haemoglobin [6,10,15]. In case of absent functional G6PD NADPH cannot be produced in sufficient amount for regeneration of glutathione resulting in missing defence against oxidative damage thus haemolysis [6,10,15].

Typical surgery

Not reported.

Type of anaesthesia

There is NO definite recommendation for either general or regional anaesthesia.

The available literature recommends avoiding drugs that cause oxidative stress and drugs that can induce methaemoglobinaemia [18]. Methylene blue treatment for methaemoglobinaemia is ineffective in G6PD deficient patients and it may also lead to severe haemolysis due to its weak oxidizing ability [17]. Therapy of methaemoglobinaemia in G6PD deficient patients consists of blood transfusions, hyperbaric oxygen therapy [17] and urine alkalinisation.

There does not exist an evidence-based global consensus regarding medication use in G6PD deficient patients [47].

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Necessary additional diagnostic procedures (preoperative)

Beside the standard preoperative investigations (evaluation of medical history, physical examination [neonatal jaundice], blood test), an extended blood testing including haptoglobin, lactate dehydrogenase, total and unconjugated bilirubin and reticulocytes can be done.

Elective surgery should not be undertaken during a haemolytic episode or in presence of an infection [25].

The patients class of G6PD deficiency should be checked to know the degree of deficiency and disease manifestation by verification of enzyme defect (reduced activity: reference range for men 2.70 – 6.62 and for women 3.25 – 7.87 U/gHb) [42] or PCR.

It can be useful to provide a list of contraindicated drugs to the medical file (or patient`s ID) of the patient as a precaution that the patient does not receive any oxidant drugs on the ward, in the recovery room, etc.

It is supposed that patients with G6PD deficiency have a lower mortality from cancer and cardiovascular diseases [13,14,30].

It is known that G6PD deficient cells provide protective effect against the malaria parasite Plasmodium falciparum in women by means of early phagocytosis of macrophages [9,41,46].

Particular preparation for airway management

Not reported.

Particular preparation for transfusion or administration of blood products

Assure the availability of cross-matched blood products to be ready in the event of significant haemolysis, especially in patients with severe enzyme deficiency (class I and class II) [41].

Particular preparation for anticoagulation

Chowdry et al., 2012, and Porto et al., 2011, administered heparin without signs of haemolysis in their case reports [12,35]. Aspirin is discussed controversially regarding safety in patients with G6PD deficiency [17] as you can see in table 3. Porto et al., 2011, report an uneventful taking of Clopidogrel regarding haemolysis [35].

Particular precautions for positioning, transport or mobilisation

Patients should have first priority in the theatre to reduce preoperative stress response. Aggressive treatment of perioperative hyperglycaemia (tight glycaemic control). Close temperature control (intraoperative use of warming blankets).

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

Patients with chronic non-spherocytic haemolytic anaemia due to class I G6PD deficiency variants may take vitamin E or folic acid (up to date). No drug interactions are reported in this regard.

Anaesthesiologic procedure

Anaesthetic management should focus on avoiding drugs that cause oxidative stress or can induce methemoglobinaemia [17,19]. Provide generously analgesia as stress related to surgery can cause haemolysis. Monitor and treat haemolysis should it occur [45].

Table 1

SUBSTANCE	SAFE
Acetaminophen (Paracetamol)	☑ 6,18,31
Amikacin	☑ 44
Bupivacaine	☑ 2,26
Chloroquine	☑ 6
Clopidogrel	☑ 34
Glycopyrrolat	☑ 44
Halothan	☑ 4,17
Heparin	☑ 12,34
Ibuprofen	☑ 31
Ketamine	☑ 4,17
Mannitol	☑ 12,44
N_2O	☑ 37,40,44
Neostigmin	☑ 44
Parecoxib	☑ 31
Pethidine	☑ 33
Phenytoin	☑ 6,17,18,46
Propofol	☑ 2,17,37
Rocuronium	☑ 44
Succinylcholine	☑ 15
Sufentanil	☑ 31
Thiopental	☑ 15,40
Tramadol	☑ 31

Table 1 contains a non-comprehensive list of frequent used drugs in the perioperative period (for anaesthesia and pain therapy) that probably can be safely given in normal therapeutic doses in patients with G6PD deficiency.

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Table 2

	CONTROVERSIALLY DISCUSSED	
SUBSTANCE	**SAFE (in normal therapeutic doses and/or in mild G6PD IV-V variants)**	**UNSAFE**
Alfentanil	☑ 33	☑ 17
Ascorbic acid	☑ 6,10,17,18,46	☑ 10,15,46*
Aspirin (low dose)	☑ 3,6,10,17,18,39,46	☑ 10,11,15,35,46*
Chloramphenicol	☑ 6,10,17,18,46	☑ 3,10,11,15,46*
Fentanyl	☑ 2,12,15,17,24,26,33,37,44	☑ 33 (?)
Glibenclamide	☑ 46	☑ 6,46*
Isoflorane	☑ 12,15,26,40,44	☑ 4,17
Isoniazid	☑ 6,17,18,46	☑ 15,46*
Metamizole	☑ 46	☑ 6,15,46*
Midazolam	☑ 2,12,17,26,44	☑ 4
Nitroprusside	☑ 34	☑ 40
Paracetamol	☑ 10,19,31,44,46	☑ 17,20,27,28,36,44,46*
Penicillin	☑ 24	☑ 15
Prilocain	☑ 17,18, 46	☑ 15
Remifentanil	☑ 33	☑ 33 (?)
Sevoflurane	☑ 26,37,44	☑ 4,17
Streptomycin	☑ 6,17,18,46	☑ 15
Trimethoprim	☑ 6,17,18,46	☑ 10, 42,46*
Vitamin K	☑ 6,10,18,46	☑ 10,15,17

Table 2 contains a non-comprehensive list of frequent used drugs in the perioperative period (for anaesthesia and pain therapy) that are controversially discussed in patients with G6PD deficiency.

*Youngster et al., 2010, in his evidence-based review stated, that these drugs have been considered unsafe, but overall evidence does not contravene use in G6PD deficient patients [46].

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Table 3

SUBSTANCE	UNSAFE
Acetanilid	☑ 6
Acetazolamid (Diamox®)	☑ 15
Aspirin (high dose)	☑ 46
Co-Trimoxazole*	☑ 10,42,46*
Dapsone	☑ 10,15,17,18,46
Diclofenac	☑ 15
Diazepam	☑ 4,15,17
Gentamicin	☑ 24
Lidocaine	☑ 15,17
Methylene blue	☑ 6,7,15,17,18,29,46
Metoclopramide	☑ 15
Naphthalene	☑ 6
Nitrofurantoin	☑ 6,10,15,17,18,46
Nitroprussidnatrium	☑ 15
Penicillin	☑ 15
Phenazopyridine	☑ 6,10,46
Prilocaine, e.g. EMLA® cream	☑ 6,16
Primaquine/ Pamaquine	☑ 3.6,10,15,17,18,46
Quinolone antibiotics, e.g. Nalidixic acid, Ciprofloxacin*	☑ 6,10,15,46*
Rasburicase	☑ 8,32,46
Sulfonamide antibiotics	☑ 3,6,10,15,17,18
Toluidine blue	☑ 6,15,17,18,46

Table 3 contains a non-comprehensive list of frequent used drugs in the perioperative period (for anaesthesia and pain therapy) that are unsafe in patients with G6PD deficiency.

*Youngster et al., 2010, in his evidence-based review stated, that these drugs have been considered unsafe, but overall evidence does not contravene use in G-6-PD deficient patients [46].

Particular or additional monitoring

Monitor temperature and check blood gases to avoid hypothermia [17] and detect acidosis and hyperglycaemia, which are potential precipitating factors for haemolysis. Keep an eye on the excreted urine to detect haemoglobinuria as a sign of active haemolysis [18].

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Possible complications

Patients with G6PD deficiency are at risk for haemolytic crisis due to exposure to oxidative stressors like certain drugs, the surgical intervention per se, metabolic conditions (metabolic acidosis, ketoacidosis), and infections [5,17,18,28,44]. The production of oxidative radicals in the case of reperfusion following ischaemia during an operation can cause haemolysis in G6PD deficiency [22, 45].

Patients with glucose-6-phosphate dehydrogenase deficiency who are undergoing cardiac surgery may have a more complicated course with a longer ventilation time, more hypoxia, increased haemolysis, and a need for more blood transfusion [16].

Younker et al. report one case of malignant hyperthermia in a patient with G6PD deficiency. However, diagnosis of malignant hyperthermia in the form of an in-vitro muscle test was not mentioned in the paper. According to our literature research, this is the only published paper to describe malignant hyperthermia in a G6PD-deficient patient [48].

Postoperative care

Clinical signs and symptoms of haemolysis typically arise within 24 to 72 hours after exposure to the triggering agent [18]. Typical laboratory workup in a haemolytic crisis reveals decreased haemoglobin and haptoglobin whereas the levels of lactate dehydrogenase, unconjugated bilirubin and reticulocytes are elevated [10,18]. Direct coombs test should be negative, because G6PD deficiency is not an immune process. In peripheral blood smear Heinz bodies, schistocytes and reticulocytes can be found. The patient should be monitored closely in a haemolytic crisis. In most cases of acute haemolysis, no specific treatment is necessary [18]. Mainly in the black population haemolysis is often self-limited as soon as the older red blood cells (< 25%), which are more deficient than the younger ones, decompose [15]. More severe diseases are seen in the white population that may result in haemoglobinuria and acute renal failure [10]. Treatment of haemolysis consists of eliminating the triggering factor and controlling the clinical symptoms. Rarely blood transfusions are required, except in children. Haemoglobin concentrations recover after 8 to 10 days [18]. If there is a severe complication its mostly acute renal failure [11,21,23], therefore renal protective measures like volume support, forced diuresis and alkalization of urine are recommended in case of haemolysis [15]. Infections and stress may precipitate haemolysis. This underlines the importance of antibiotic treatment in time, adequate analgesia and anxiolysis in the perioperative period [45].

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 disease

Consider that in a narcotized patient it is not possible to identify immediate clinical signs of haemolysis like fatigue, back pain, headache and dyspnoea while the patient is asleep. Hypotension or tachycardia, which can be a sign of haemolysis, may be attributed to other causes in a patient under general anaesthesia. Further signs of haemolysis such as cyanosis, jaundice, renal failure, dark urine und splenomegaly will usually arise within 24 to 72 hours after exposure to the triggering agent [18].

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Ambulatory anaesthesia

The patient should be informed about the signs and symptoms of haemolysis, which usually arise within 24 to 72 hours after exposure to the triggering agent [18]. For this reason, ambulatory anaesthesia is not recommended.

Obstetrical anaesthesia

The gene for G6PD deficiency is located on the X chromosome and inherited in an X-linked fashion, being fully expressed in hemizygous males and homozygous females, but in only a proportion of female heterozygotes [46]. The enzyme activity of heterozygous females may be normal, moderately reduced or severely deficient, depending on the degree of X chromosome inacitivation (lyonisation) [456].

It is hypothesized that pregnant women with G6PD deficiency are at greater risk for development of preeclampsia than are those with normal G6PD activity [1]. Furthermore, increased rates of abortion, low birthweight infants, and puerperal drops in red cell volumes in G6PD-deficient pregnant women were noted [34].

Neonatal jaundice is a serious complication of G6PD deficiency occurring during the first week of life since bilirubin-induced neurotoxicity can lead to severe neurologic consequences (kernicterus) [16,49,50].

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These guidelines have been prepared by:

Authors
Agnes Foedinger, Anaesthesiologist, University Hospital Innsbruck, Austria
agnes.foedinger@uki.at

Thomas J. Luger, Anaesthesiologist, University Hospital Innsbruck, Austria
thomas.luger@i-med.ac.at

Peer Revision 1
Gabor Erdoes, Anaesthesiologist, University Hospital Inselspital Bern, Switzerland
gabor.erdoes@insel.ch

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Lucio Luzzatto, Scientific Director Istituto Toscano Tumori, Firenze, Italy
lucio.luzzatto@ittumori.it

C.J.F. Van Noorden, Academic Medical Center, Head of department Cell Biology & Histology, University of Amsterdam, The Netherlands
c.j.vannoorden@amc.uva.n

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