International Journal of Scientific Reports
Munasinghe BM et al. Int J Sci Rep. 2020 Dec;6(12):526-528
http://www.sci-rep.com
pISSN 2454-2156 | eISSN 2454-2164
DOI: https://dx.doi.org/10.18203/issn.2454-2156.IntJSciRep20205032

Case Report

Anaesthesia for a child with glucose-6-phosphate-dehydrogenase deficiency: a case report

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Received: 10 October 2020
Accepted: 11 November 2020

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ABSTRACT

Patients with glucose-6-phosphate-dehydrogenase (G6PD) deficiency are prone to acute haemolytic crisis during perioperative period, precipitated by oxidative stress and certain medications, which are common concurrences during this period. Careful peri-operative planning, judicious use of medication and postoperative monitoring plays a pivotal role in preventing complications in these patients. Here, we report a successful case of general anaesthesia in a child with G6PD deficiency.

Keywords: Glucose-6-phosphate dehydrogenase deficiency, Oxidative stress, Acute haemolytic crisis

INTRODUCTION

G6PD deficiency, the commonest red cell enzymopathy in humans, affects around 400 million people on the globe.[1] It is a X-linked recessive disorder, triggering a defective pentose phosphate pathway (PPP) making red cells vulnerable to oxidative stresses. The affected red cells in such a background are prone to acute haemolytic crisis. Perioperative period carries increased risk of acute haemolytic crisis due to the use of certain drugs, surgical stress and infections, as all these increase the oxidative burden.

CASE REPORT

A 5-year-old, 15 kg child was scheduled for surgical correction of congenital talipes equinovarus. He had been diagnosed with G6PD deficiency after being investigated for neonatal jaundice, indirect hyperbilirubinaemia and anaemia with a blood picture suggestive of oxidative-stress-induced haemolysis. He had required blood transfusion once but no other episodes of acute or chronic haemolysis were noted. However, he had neurocognitive delay. At pre-operative visit, he was neither pale nor jaundiced. The airway assessment and rest of the systemic examination were normal. The echocardiogram revealed an uncomplicated atrial septal defect. Haemoglobin was 12.9 g/dl with a reticulocyte count of 1.1% (normal- 0.82-1.45). White cell and platelet counts, total and indirect bilirubin levels and serum potassium were all within normal range. Haematology review was arranged, blood was grouped and cross matched. Informed written consent was taken from the parents. The child was fasted 6 hours for solids and 2 hours for clear liquids. Syrup paracetamol 225 mg was given 1 hour prior to the surgery. At operating theatre, standard monitoring was established including ECG, non-invasive blood pressure and peripheral saturation. A 22 G cannula was inserted and a crystalloid drip commenced. Intravenous co-amoxiclav 450 mg was administered as surgical prophylaxis. The child remained calm throughout this period and anaesthesia was induced with intravenous propofol 40 mg, fentanyl 30 µg and atracurium 8 mg. Airway was secured with a size 5.5 mm ET tube. Urinary catheter was inserted. Caudal block was

administered with 12 ml of 0.25% plain bupivacaine. Patient warmer was applied and nasopharyngeal temperature monitoring initiated. With regard to the use of tourniquet, collective decision was made to utilize it only in case of significant bleeding. Anaesthesia was maintained with isoflurane, O^2 and N^2O. Fine surgical dissection was performed, while close monitoring for blood loss. Child remained stable with normal core temperature, end tidal CO^2 levels, capillary blood sugar, adequate urine output and normal urine colour. Blood loss was less than 40 ml. Residual paralysis was reversed at the end of the 2 hours surgery. Child was extubated and sent to ward for observation. Post-operatively, subcutaneous morphine 1 mg 6 hourly and regular doses of oral paracetamol were provided for analgesia. Oozing was noticed from surgical site 6 hours later which settled gradually. Distal perfusion of the limb was satisfactory. Repeated haemoglobin level was 12.6 g/dl. Blood picture, serum bilirubin levels and reticulocyte count done the next day did not reveal any haemolysis. Pain was minimal. The child was closely monitored during the next two days and sent home 72 hours later with instructions to parents to seek medical advice with any sign of infection or haemolytic crisis. He had an uneventful post-operative period and reviewed in the orthopaedic clinic in 2 weeks.

DISCUSSION

G6PD catalyses formation of NADPH in PPP. This in turn aids replenishing antioxidant glutathione, which is vital for red cells in combating oxidative stresses. Deficiency of G6PD could therefore lead to denaturing of haemoglobin and haemolysis.[1] Fava beans, infection, certain drugs and metabolic acidosis are common precipitants of haemolysis.[2]

Certain classes of antibiotics (dapsone, fluoroquinolones, nitrofurantoin etc.), Methylene blue and anti- malarials are well-known medications implicated in acute haemolytic crisis in G6PD patients. With regard to anaesthetic drugs, isoflurane, sevoflurane and benzodiazepines have been found to have inhibitory effects on G6PD activity in vitro.[3] However, several case reports suggest their safe use during anaesthesia.[4-6] In our patient, use of isoflurane did not result in haemolysis. Sharma et al, reported use of total intravenous anaesthesia with propofol, remifentanil and dexmeditomidine.[7] The former agents are considered to have antioxidant properties thus may have protective properties when used in this group of patients. In several cases, bupivacaine has been used without increased risk of haemolysis.[7,8] NADPH which is deficient in these patients is useful to reduce methemoglobin, an ineffective variant of haemoglobin. Moreover, methylene blue used in the treatment of methemoglobinaemia, is not efficacious in G6PD deficient patients. Thus, the current guidance advocates against the use of lignocaine and prilocaine (which could occasionally precipitate methemoglobinaemia).[9] Thus, EMLA was not used for topical anaesthesia prior to cannulation in this child but we performed a smooth cannulation and induction.

Bhaskaran et al reported a case where use of tourniquet resulted in acute heamolytic crisis in a G6PD deficient adult during a lower limb orthopaedic surgery.[10] This was postulated to be due to distal ischaemia, reperfusion and metabolic acidosis up to now. No clear guidance exists regarding the use of tourniquet in this group of patients. On the other hand, the common factors such as intraoperative hypothermia, hypoxia, acidosis, poor glycemic control, infection and surgical stress have been clearly associated with haemolytic crisis thus should be obviated. These factors were optimized in our child while careful monitoring for acute haemolysis was continued by observing for pallor, altered colour of urine (hemoglobinuria), drop in haemoglobin and rise in reticulocyte count and indirect bilirubin level.

The management of pain is pertinent in reducing stress response and should be multimodal. Regional nerve blocks or neuraxial blocks could be utilized for both surgical anesthesia and post-operative analgesia as well. Use of paracetamol is controversial in this aspect where some suggest controlled use while avoiding overdose NSAIDs and opiates could be used depending on the intensity of pain. As this child did not have history of haemolysis with paracetamol, it prompted us its use in conjunction with morphine.[1,11]

Haemolysis in G6PD patients typically occurs within 24-72 hours after exposure to a stressor. Thus, it is extremely important to continue monitoring.[1] There could be pallor, jaundice, headache and haemoglobinuria. Blood picture would show Heinz bodies. Serum lactate dehydrogenase levels will be elevated while serum haptoglobin is reduced. In a case of a haemolytic episode, trigger should be removed. Patient should be adequately hydrated.[7] Diuretics could be used to promote urine output. Blood transfusions may be needed only infrequently.[5] Such episodes usually settle by day 10.[1] In our patient, careful planning and monitoring with avoidance of triggers led to an uncomplicated recovery.

CONCLUSION

Anaesthesia for G6PD deficient patients presents a unique challenge to anaesthetists. Controversies regarding frequently used anaesthetic drugs (such as benzodiazepines, inhalational agents, analgesics) and tourniquet warrant further studies and data. The priority would be to avoid oxidative stressors, monitor, detect and manage adverse events promptly, that would lead to uncomplicated recovery following surgery.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Munasinghe BM, Nimalan S, Subramaniam N, Goonetilleke S. Anaesthesia for a child with glucose-6-phosphate-dehydrogenase deficiency: a case report. Int J Sci Rep 2020;6(12):526-8.