Anaesthesia for a patient with B-thalassaemia

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B-Thalassaemia is a rare hereditary disease caused by partial or complete deficiency of β-haemoglobin chain synthesis. There is a lot of literature regarding anaesthetic management in other haemoglobinopathies (i.e. sickle cell disease), especially in the paediatric population, but there is scarce information regarding β-thalassaemia major in adults. With current medical management, β-thalassaemia major patients survive to adulthood and may present for a variety of surgical procedures, even unrelated to their disease process. It is important for the anaesthetist to be familiar with the pathophysiology of β-thalassaemia major and how the disease itself and its treatment or complications (iron deposition from multiple transfusions) can affect anaesthesia. A case of a 51-year-old woman with severe β-thalassaemia undergoing laparoscopic cholecystectomy is presented. The anaesthetic management and systematic review of the perioperative concerns in severe β-thalassaemia are discussed.

Keywords: anaesthesia, B-thalassaemia, laparoscopic cholecystectomy, non-cardiac surgery, pathophysiology

Case report

A 51-year-old woman with β-thalassaemia, of American Society of Anesthesiologists (ASA) physical status IV, presented for laparoscopic cholecystectomy. Her past medical history was significant for severe anaemia requiring multiple red blood cell transfusions since early childhood (on average two units of washed red blood cells every two weeks for a haemoglobin of less than 8.5 g/dl). This led to severe iron deposition (haemosiderosis) with severe multiple organ insufficiency. She suffered from right-sided heart failure with lower extremities oedema and ascites. Over the last year, she had had recurrent episodes of paroxysmal atrial fibrillation and was on oral anticoagulation and digoxin. She was diagnosed with pulmonary hypertension and her exercise tolerance was less than 10 uphill steps. She had cirrhosis Child–Pugh class B. She had hypothyroidism, primary ovarian failure and osteoporosis. Her last transfusion was a few days prior to surgery. She had a splenectomy at the age of 11.

Her home medications included digoxin, spironolactone, furosemide 80 to 125 mg on alternate days, thyroxine, ursodeoxycholic acid, allopurinol, deferiprone (oral chelating agent), calcium and warfarin (changed to low molecular weight heparin (LMWH) perioperatively).

On physical examination the patient was thin (41 kg). Her airway assessment revealed a Mallampati score of II, 4 cm mouth opening, 5 cm thyromental distance, short neck and tonsillar enlargement. She had frontal and maxillary bossing but no significant mandibular enlargement. On auscultation she was in a regular rhythm with an S3 sound. Her lower extremities were oedematous and ascites was present. She had no secondary sexual characteristics (pubic/axillary hair, breasts).

Her pertinent laboratory values were (abnormal values are given in bold):

<table>
<thead>
<tr>
<th>Laboratory tests</th>
<th>Patient’s values</th>
<th>Normal values (SI units)</th>
</tr>
</thead>
<tbody>
<tr>
<td>White cell count (WCC)</td>
<td>16.2  c/mm³</td>
<td>4.9-10.8  c/mm³</td>
</tr>
<tr>
<td>Haemoglobin (Hb)</td>
<td>12.1  g/dl</td>
<td>11.8-17.8  g/dl</td>
</tr>
<tr>
<td>Haematocrit (Hct)</td>
<td>36.8%</td>
<td>37.4-47.9%</td>
</tr>
<tr>
<td>Platelets (PLTs)</td>
<td>415  c/mm³</td>
<td>150-350  c/mm³</td>
</tr>
<tr>
<td>Prothrombin time (PT)</td>
<td>11.2 s</td>
<td>11-13.5 s</td>
</tr>
<tr>
<td>Sodium (Na)</td>
<td>130 mEq/L</td>
<td>136-150 mEq/L</td>
</tr>
<tr>
<td>Potassium (K)</td>
<td>4.9 mEq/L</td>
<td>3.5-5.5 mEq/L</td>
</tr>
<tr>
<td>Blood urea nitrogen (BUN)</td>
<td>51.05 mmol/L</td>
<td>8.0-16.4 mmol/L</td>
</tr>
<tr>
<td>Creatinine (Cr)</td>
<td>88.4 micromol/L</td>
<td>50-110 micromol/L</td>
</tr>
<tr>
<td>Thyroid stimulating hormone (TSH)</td>
<td>2.74 mU/L</td>
<td>0.4-4 mU/L</td>
</tr>
<tr>
<td>Total bilirubin (T. Bil)</td>
<td>1.8 mg/dl</td>
<td>0.20-1.2 mg/dl</td>
</tr>
<tr>
<td>Direct bilirubin (B. Bil)</td>
<td>0.8 mg/dl</td>
<td>0.0-0.5 mg/dl</td>
</tr>
<tr>
<td>Transaminase (AST)</td>
<td>37 IU/L</td>
<td>0-45 IU/L</td>
</tr>
<tr>
<td>Transaminase (ALT)</td>
<td>30 IU/L</td>
<td>0-45 IU/L</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>79 IU/L</td>
<td>40-150 IU/L</td>
</tr>
</tbody>
</table>

The electrocardiogram (ECG) showed sinus tachycardia at 98 beats per minute (bpm). Echocardiography revealed a dilated right atrium and right ventricle, decreased right ventricular function, mild mitral regurgitation and pulmonary hypertension. The left ventricle was not dilated and had good function. A chest radiograph showed cardiomegaly and an increased cardiothoracic ratio.

On arrival in the operating theatre standard ASA monitors, with the addition of an awake arterial line, were placed. The patient’s blood pressure was 80/55 mm Hg with sinus rhythm at 80 bpm, while arterial haemoglobin oxygen saturation (SpO₂) was 97% at room air. A smooth intravenous induction was performed with etomidate, lidocaine and fentanyl. Paralysis was achieved with
rocuronium. Despite the facial deformity, tracheal intubation was uneventful. Anaesthesia was maintained with sevoflurane, O₂ and intermittent fentanyl boluses. Peritoneal insufflation with CO₂ was performed slowly, with the pressure limit set at 12 mm Hg. Throughout the operation vital signs were stable and oxygenation was followed with serial arterial blood gases (ABGs). The operation lasted approximately 40 min. At the conclusion of surgery residual paralysis was reversed with sugammadex and the patient was extubated after regaining consciousness. A total of 200 ml 0.9% normal saline (NS) was administered during the procedure (0.9% NS was preferred to other crystalloid solutions for its low potassium content). Postoperative pain was controlled with intravenous paracetamol. No anaesthetic or surgical complications were noted. She was discharged from hospital the following day.

Discussion
B-thalassaemia is a congenital haemolytic disorder caused by partial or complete deficiency of β-globin chain synthesis. It primarily affects people of Mediterranean origin and to a lesser extent people of Chinese, Asian and African descent. It is usually described in its mild form, B-thalassaemia intermedia. These patients are anaemic but do not require frequent transfusions. They may have hepatosplenomegaly. B-thalassaemia major arises from the absence of synthesis of β-globin chains (β-0 thal). It is inherited on chromosome 11 and patients are homozygous for two mutant alleles. Anaesthetic management of these patients is dictated by the disease itself, the complications of its treatment (iron overload from multiple transfusions) and the use of preventive measures (chelating agents).

Anaesthetic considerations
Airway
B-thalassaemia major produces maximum erythropoietin release and massive marrow hyperplasia. Patients may present with frontal bossing and expansion of the maxillofacial area. There is a possibility of difficult mask ventilation and intubation due to facial deformity. Extramedullary haematopoiesis may also occur in the vertebral canal, compressing the spinal cord and causing several neurologic symptoms, necessitating caution during neck manipulation at laryngoscopy. Abnormal dentition increases the intubation difficulty and the possibility of tooth damage during intubation attempts.

Cardiovascular
Cardiac insufficiency, secondary to haemosiderosis from chronic transfusions, ranging from dyspnoea and peripheral oedema to congestive cardiomyopathy and cardiac failure may be present. Major β-thalassaemia patients may present with pulmonary hypertension and vascular damage (vasculopathy), secondary to disturbed nitric oxide physiology. Angina, conduction defects, pericarditis, arrhythmias and sudden death are all more likely in patients with thalassaemia and iron overload (> 2500 µg/L serum ferritin). A hyper-dynamic circulation with a high cardiac output, a compensatory mechanism to chronic anaemia, and decreased blood oxygen carrying capacity may also be present.

Liver
Chronic anaemia induces ineffective erythropoiesis in extramedullary erythropoietic tissue. Hepatomegaly may be present. Haemochromatosis, due to chronic transfusion therapy, can lead to liver fibrosis, cirrhosis, hepatic failure and hepatocellular carcinoma. Liver failure decreases the synthesis of plasma-binding proteins and causes hypoalbuminaemia.

Haematological
Chronic haemolysis leads to chronic anaemia and hyperbiliru-
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 nevertheless, it was not until her twenties that iron chelation therapy was introduced. Our patient exhibits an interesting pathophysiology, as she survived past childhood due to the initiation of chronic hyper-transfusion therapy, but suffered almost all of the complications of iron overload due to delay in the initiation of chelating therapy in the 1960s when she was born.

The anaesthetic management of this severe form of β-thalassaemia requires multiple considerations including the airway, cardiovascular system, liver and coagulation system. Anaesthetists should be aware of both the pathophysiology of the disease and the potential complications of its treatment. Meticulous consideration should be paid to each one of the systems involved in order to achieve the best possible outcome in these patients.

References

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