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Preoperative assessment of patients with osteogenesis imperfecta

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Osteogenesis imperfecta (OI) is an inherited disorder of the connective tissue the primary manifestation of which is an increased susceptibility to fractures. Patients often require orthopaedic treatment of fractures and resultant deformities. Commonly encountered intraoperative complications include difficult airway with easily fractured teeth, patient positioning problems, intraoperative bleeding, hypothermia, respiratory compromise due to skeletal deformity, cardiac problems or basilar invagination caused by congenital anomalies, and very rarely development of perioperative hyperthermia. Therefore, multidisciplinary approach to preoperative assessment of patients with OI, including identification of risk factors and optimization of health before surgery is needed.

Key words: intraoperative complications; osteogenesis imperfecta; preoperative assessment

INTRODUCTION

Osteogenesis Imperfecta (OI) is an inherited disorder of the connective tissue the primary manifestation of which is an increased susceptibility to fractures. Commonly encountered intraoperative complications include difficult airway with easily fractured teeth, patient positioning problems, intraoperative bleeding (coagulation or platelet dysfunction and technical aspect of the surgery), increased tendency to hypothermia due to the duration of surgery, respiratory compromise due to skeletal deformity, and the last but not the least, cardiac problems caused by congenital anomalies, basilar invagination, and very rarely development of perioperative hyperthermia.

Proper preoperative assessment, correct identification of risk factors, and optimization of health before surgery is important, as is the choice of anaesthetic technique.

PROCEDURE RISK LEVEL

Surgical placement of simple intramedullary rods to prevent or correct long-bone deformities are usually low to moderate risk procedures, but placement of telescoping rods, which may be appropriate for patients older than two years who are actively growing, is usually long-lasting and characterized by a significant loss of blood. These are high-risk procedures.

PATIENT RELATED RISK FACTORS

In OI patients, most of the interventions are usually performed at the age of two to ten years and they are usually classified as ASA score 1-2. The main and most common clinical manifestations of OI important for anaesthesia are short stature, scoliosis, megalocephaly, short neck, limited range of motion of cervical spine, basilar skull deformities that may cause nerve compression or other neurologic symptoms, hearing loss, opalescent fragile teeth, easy bruising, bone fragility, and joint laxity.

Good physical examination includes examination of the oral cavity and the teeth, Mallampati score, hearing check, examination of tongue size, neck and facial deformity, neck mobility, thyromental distance, central venous access, auscultation of the lungs and the heart, and finally, access to peripheral blood vessels. It is mandatory to check the lumbar region due epidural catheter setting. However, spinal deformity associated with OI may make neuraxial anaesthe-

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sia (i.e. epidural, spinal, or combined spinal epidural) technically challenging and may make the spread of neuraxial local anaesthetic unpredictable.

Patients with OI are at risk of facial bone fractures, cervical spine injury, and dental damage during endotracheal intubation, placement of supraglottic airways, or even mask ventilation. They are at risk of positioning-related fracture and joint dislocation. Fracture or bruising may also occur with excessive inflation of the blood pressure cuff.

The cardiopulmonary abnormalities that are associated with OI (i.e. kyphoscoliosis, restrictive lung disease, valvular heart disease, aortic root dilatation) may require invasive monitoring and/or modification of the choice of anaesthetic induction agents. Patients with type III deforming OI or other moderate to severe types of OI should have yearly spirometry (to evaluate for restrictive lung disease due to kyphoscoliosis and chest deformities) and an electrocardiogram (ECG) and echocardiogram (cardiac echo) every two years to detect aortic root dilation and valvular dysfunction. Chest and spine radiography performed every one to two years are usually helpful, but it is recommended to perform cardiac echo for up to six months, along with follow-up spirometry. ECG and cardiac echo of the heart and spirometry also include the opinion of competent paediatricians.

Basilar skull deformity may lead to basilar invagination. Thus, neurologic examination and magnetic resonance imaging/multi-slice computed tomography should be performed as indicated by symptoms or behavioural changes, particularly in patients with type III OI.

Bleeding diathesis related to capillary fragility and platelet function abnormalities is associated with OI and can result in significant haemorrhage during surgery or slow oozing after the procedure. Long-lasting treatments are characterized by bleeding from the bone canal and suppressing the extremities, which also increases blood loss. Bleeding time and standard coagulation tests including prothrombin time, INR, activated partial thromboplastin time, fibrinogen level, d-dimer, and platelet number with platelet aggregation should be obtained prior to any surgical procedure. If there is a history suggestive of abnormal bleeding, haematologist and/or transfusiologist consultation is advised. Nonsteroidal anti-inflammatory drugs should be avoided because of their antiplatelet effects.

Hyperthermia may occur in patients with OI for unclear reasons and the mechanism has been linked to either malignant hyperthermia or increased thyroid hormones, but most of these patients are not at an increased risk of malignant hyperthermia.

Typical laboratory findings in patients with OI are elevated levels of serum alkaline phosphatase, hypercalciuria, and hypercalcemia after bisphosphonate therapy. A common but not typical finding is iron deficiency anaemia due to social reasons and eating habits. Reduced levels of protein and fibrinogen are not uncommon. A routine preoperative laboratory examination should include complete blood count, electrolyte levels (especially calcium), liver and kidney function tests, and complete coagulogram. In case of hyperthyroidism, the concentrations of triiodothyronine, thyroxine and thyroid-stimulating hormone should be analysed, but these tests should not be older than six months. Endocrinologist opinion should also be obtained.

To reduce the risk of osteomyelitis occurrence, it is necessary to obtain swab of the nose, throat and skin.

CONCLUSION

Many anaesthetic problems may be encountered in orthopaedic treatment of deformity in patients with OI, caused by positioning the patient on the operating table, malignant hyperthermia, basilar invagination, or cardiac problems due to congenital anomalies. However, the most common are difficult airway, fragile teeth, central venous or epidural catheter placing problems, intraoperative bleeding with fluid shift and consequent coagulation disorder, hypothermia, and finally possible postoperative respiratory function problems. Proper preparation, optimization of health before surgery, and preoperative assessment are important. This implies a multidisciplinary approach to patients with OI that includes different physicians such as orthopaedist, paediatrician, transfusiologist, radiologist, operating room staff, psychologist, etc., which should lead to an uneventful anaesthetic course.

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SAŽETAK

Prijeoperacijska procjena bolesnika s osteogenesis imperfecta

Slobodan Mićić, Goran Pavelić, Tomislav Čutura, Ana Blagaić

Osteogenesis imperfecta (OI) je nasljedna bolest vezivnog tkiva, a njezina glavna manifestacija je povećana sklonost prijelomima. Ovi bolesnici često zahtijevaju ortopedsko liječenje prijeloma i posljedičnih deformiteta. Česte intraoperacijske komplikacije su otežani dišni put s lako lomljivim zubima, problemi pozicioniranja bolesnika, intraoperacijsko krvarenje, hipotermija, respiracijski poremećaji zbog koštanih deformacija, srčani problemi ili bazilarna invaginacija uzrokovana prirođenim anomalijama te vrlo rijetko razvoj perioperacijske hipertermije. Stoga je neophodan multidisciplinski pristup prijeoperacijskoj procjeni bolesnika s OI, uključujući utvrđivanje rizičnih čimbenika i postizanje optimalnog zdravstvenog stanja prije operacije.

Ključne riječi: intraoperacijske komplikacije; osteogenesis imperfecta; prijeoperacijska procjena