

## Monitored Anesthesia Care in Dental Treatment of a Patient With Laurence-Moon-Bardet-Biedl Syndrome

Laurence-Moon-Bardet-Biedl Sendromlu Bir Hastanın Dental Tedavisinde Monitörize Anestezi Bakımı

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Laurence-Moon-Bardet-Biedl syndrome (LMBBS) is an autosomal recessive disorder characterized by cone-rod dystrophy, postaxial polydactily, central obesity, cognitive impairment, hypogonadism, renal dysfunction, retinal pigmentation degeneration, mental retardation and short stature. As the secondary features of LMBBS, heart defects, diabetes mellitus, hypothyroidism, renal diseases, hepatic, dental and facial abnormalities have been reported (1-4).

In this report, we aimed to present our anesthetic management protocol in dental treatment of a patient who has both cardinal features of LMBBS and chronic hepatic failure and hypothyroidism as co-existing disorders.

A 22-year old female patient with LMBBS has been referred to us because of her dental problems. Clinical and radiological investigation of the patient had not been done because of mental retardation and we have been consulted to evaluate and treat her under general anesthesia. Poor coordination, behavioral problems, speech problems, irritability and anxiety were evident. Macroglossia, petechias on extremities, restriction in thyromental distance, manifestation of occiput, restriction of flexion of head were observed. Airway assessment revealed a class III Mallampati airway. In laboratory examinations, TSH was 6.9 mIU/mL and platelet was 93.000/µL. Under these circumstances, because of the difficulties with intubation and saving the airway patency, monitored anesthesia care (MAC) was planned for dental examination and treatment. Because of the chronic hepatic failure and the risk of bleeding in case of dental extraction, vitamin K (Libavit K®) replacement was planned a day before the dental procedures. After a routine preoperative preparation had been completed, topical anesthetic cream (Emla®) was applied on dorsal side of right hand and peripheral vascular cannula.

The patient was medicated by intravenous (iv) 1 mg/kg bolus dose propofol (Propofol®) for the purpose of sedation followed by 3 mg/kg/hour. Monitoring included pulse oximeter for obtaining peripheral blood mean oxygen saturation (SpO<sub>2</sub>), electrocardiography, non-invasive blood pressure (NIBP) [Omni-Trak Noninvasive Vital Sign Monitoring System MR].

During the operation, the patient has been ventilated by O<sub>2</sub> at a level of 4 liter/min by nasal cannula while analgesia was maintained with remifentanyl 0.05 µg/kg/min by infusion. The patients' sedation level was aimed at 3 by Observer's Assessment of Alertness/Sedation (OAA/S).

When the sedation score was 3, enough mouth opening could be provided for dental examination and the planned treatment. Articaine-adrenaline including local anesthetic (Ultracaine DS®) was administered by infiltration anesthesia to provide postoperative anesthesia and reduce bleeding at the extraction sites. During the operation, the values of SpO<sub>2</sub>, heart rates and NIBP were %97-100, 63-85 beats/min, 105-85/55-40 mmHg (respectively). Infusions were ended when the dental procedures completed. Three minutes after the infusion of medications, OAA/S score of the patient was 5. The patient was stable and taken to service.

Patients with LMBBS frequently require multiple anesthetic procedures for both diagnostic and therapeutic measures (2). MAC was preferred due to difficulties in intubation and surgical procedures caused by restriction of mouth opening (5). Appropriate medications for hepatic problems and associated bleeding were administered. OAA/S score of 3 has provided us to implement favorable mouth dental procedures. Early recovery and discharge could be provided.

### Conflict of Interest

No conflict of interest was declared by the authors.

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