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Anesthetic approach to the patient withLujan-Fryns syndrome

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Introduction: Lujan-Fryns Syndrome (marfanoid habitus syndrome X-linked mental retardation) involves especially the male sex. Its frequency is unknown. Mild-moderate mental retardation, marked facial dysmoprhism, marfanoid appearance, extremities in the shape of long cylinders, and behavioral problems are characteristics of the syndrome. In this report, anesthesia approach to a case with, Lujan-FrynsSyndrome is presented.

Case: A male patient who is 30 month old and with weight of 9kg was planned to undergo operation by ear nose throat clinic for insertion of bilateral ear tube. In pediatric examination, mental retardation, extremity abnormalities, oropharynx palate abnormalities, and horse shoe kidney were found. In physical examination, the traces of previous operations he underwent for morgagnia hernia and cleft palate were observed. No pathology was established in routine biochemical and hematological investigations. Vital findings were stable. Patient underwent standard monitorization with ECG,heart rate, and pulse oximeter. Prior to induction of anesthesia. SpO2 was 90%, heart rate 105 beat/min., and blood pressure 95/47mmHg. Considering that opening air way may be difficult due to syndromic typical facial appearance, face masks, airways and laryngeal masks at varying size were prepared. For laryngoscopy, different size blades and endotracheal tubes were kept ready and premedication was not administered. Following about 5 minutes of preoxygenation, anesthesia induction was made with 3mg/kg propofol and no problem was encountered with ventilation using face mask. Mask ventilation was observed to be comfortable and following adequate muscular relaxation with sevoflurane 1,5 no I-gel was placed without using neuro muscular blockers. Maintenance anesthesia was carried out with a 2% sevoflurane, 50% azotprotokside and 50% oxygen mixture. The patients was extubated without any problems after the operation lasting about 20 minutes and after monitorization for 30 minutes in recovery room, patients was sent to his clinic.

Discussion: Diaphgram hernia and pulmonary hypoplasia are the most common anomalies occurring in this syndrome. Limited pulmonary reserves and marked facial dysmorphism makes it difficult to have a secure airway. Cardiovascular malformations should also be taken into consideration. In the present case, in spite of probable difficulty in airway and hemostability, sevoflurane-N2O maintenance enabled safe and reliable anesthesia.

Conclusion: It is our opinion that, in Lujan-Fryns syndrome, complications may decrease by a careful preopartaive evaluation, securing air way and providing optimal conditions in the induction and maintenance of anesthesia.

Biography

Ozkan Onal has completed his medical education at Gazi University Medical Faculty and he was specialized in anesthesiology in Hacettepe University Medical Faculty. He has more than 15 publications in reputed journals in the field of anesthesia.

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