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# General anaesthesia in a patient with Edwards Syndrome: A Case Report

Edwards Sendromlu Hastada Genel Anestezi: Olgu Sunumu

Anesteziyoloji ve Reanimasyon

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## Özet

Edwards sendromu (ES), 8000 canlı doğumda bir görülen 18. kromozomdaki malformasyonlardan kaynaklanan ciddi ölümcül anomalilerle seyreden bir sendromdur. Erkek/kız oranı 1/4'tür. Doğmuş bebeklerin yüzde doksanı ilk on iki ayda kaybedilir. Hastalarda orta derecede mental retardasyon, kaslarda hipotoni ve ekstremite anomalileri, mikrosefali, fasiyal anomaliler, gelişimsel bozukluklar, skolyoz, epilepsi ve ciddi kardiyak anomaliler ve ürogenital anomaliler vardır . ES ile ilgili sınırlı sayıda literatürde, tüm olgular acil durumlarda ve ölümcül nedenleri dışlamak için anestezi almışlardır. Bu vakayı sunma amacımız, çoğu hayatının çok erken dönemlerinde kaybedilen bu sendromik hastalardan farklı olarak 13 yaşına kadar yaşamış olan ve elektif ve sorunsuz bir operasyon geçiren hastadaki genel anestezi uygulama deneyimini literatürde eşliğinde tartışmaktır.

**Anahtar kelimeler:** *Edwards Sendromu*, *Genel Anestezi*, *Dental Tedavi*

## Abstract

Edwards syndrome (ES) is a syndrome with severe fatal anomalies resulting from malformations of the 18th chromosome seen in 8000 live births. The male / female ratio is 1/4. Ninety percent of newborn babies die in the first twelve months. Patients have moderate mental retardation, muscle hypotonia and extremity anomalies, microcephaly, facial anomalies, developmental disorders, scoliosis, epilepsy, severe cardiac and urogenital anomalies. In a limited number of literature on ES, all cases were anesthetized to exclude lethal causes and emergencies. Our aim in presenting this case is to discuss the experience of anesthetic management of a 13-year- old patient with ES. He has undergone an elective and trouble-free operation and his dental treatments were successfully completed.

**Keywords:** *Edwards Syndrome*, *General Anaesthesia*, *Dental Treatment*

## Introduction

Edwards syndrome (ES) is a syndrome associated with severe fatal anomalies resulting from malformations in the 18th chromosome, seen in 1/ 8000 live births. The ratio of male to female is 1/4. Ninety percent of the babies born are lost in the first twelve months [1]. Patients have moderate mental retardation, muscular hypotonia, extremity anomalies, microcephaly, facial anomalies, developmental failures, scoliosis, epilepsy, serious cardiac anomalies and urogenital anomalies [2]. In a limited number of literature on ES, all cases were anesthetized in urgent conditions, in order to rule out the fatal causes. Our goal in presenting this case is to share with you the experience of anesthetic management of a 13-year-old patient with ES. patientwhich is passed through the elective operation in advanced age and the problem of the anesthesia which is due to the drugs and the conditions used in the literature.

## Case Report

A 13-year-old-boy weighing 73 kg was admitted to the polyclinic because of his toothache. At 2 years old, he was diagnosed with ES. 18th chromosome as short-choked microbes was confirmed genetically. Serious

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laryngospasm developing against radiopaque material given when brain MRI is withdrawn at 2.5 years, In the examination of the patient, he had dysmorphic facial appearance, and jaw and tooth structure was anatomically different. There were speech disturbance at advanced level and moderate mental retardation. There was a murmur in the mitral focus. He had Hb: 11 mg $dl^{-1}$. On echocardiogram, patent foramen ovale, pulmonary stenosis, grade I mitral insufficiency, right renal atrophy with frequent infection due to athrophic kidney were detected. He was evaluated as ASA III and mallampati 2. On the operation day, the patient was premedicated with oral midazolam 5 mg. Standard monitorization was performed. The pulse was recorded as 90 beats/min, TA 100/70 mmHg, $SPO_2$ 99% and the measurement continued at 5 minute intervals. The patient without vascular access was given induction of inhalation with 8% sevoflurane + 100% $O_2$. 22 G intravenous cannula was inserted, and 2 mg $kg^{-1}$ propofol and 0.5 mg $kg^{-1}$ rocuronium bromide were given. When respiratory depression was present, the mask was ventilated for 3 minutes with 6 lt $min^{-1}$ $O_2$ via balloon. After satisfactory muscle relaxation was achieved, portex 7 cuffed endotracheal tube intubation was performed ( Figure 1).

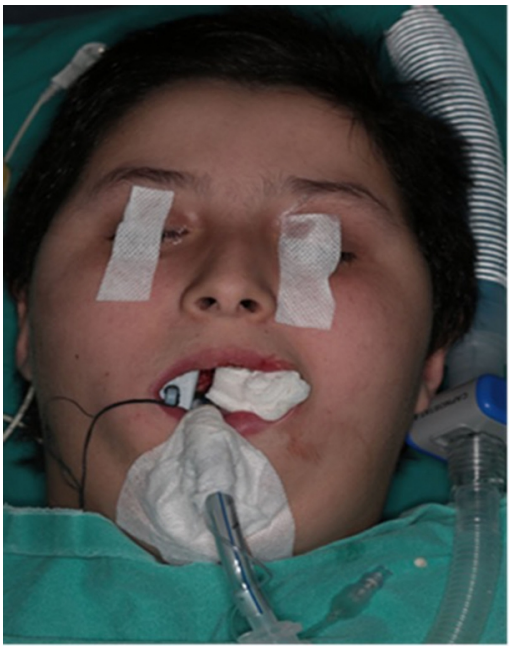

**Figure 1**
Shows the patient with Edwards syndrome under general anesthesia

After intubation, laryngospasm developed and the patient was treated with prednisolone 1mg $kg^{-1}$. %40 O2-%60 N2O- 2% anesthesia was maintained with sevoflurane. Following local infiltration anesthesia, the patient's teeth numbered 12, 16, 24, 26, 46 were filled. The patient received prophylaxis of 2gr penciline+sulbactam. Thirty minutes after this procedure, tooth 25 was extracted (Figure2-3).

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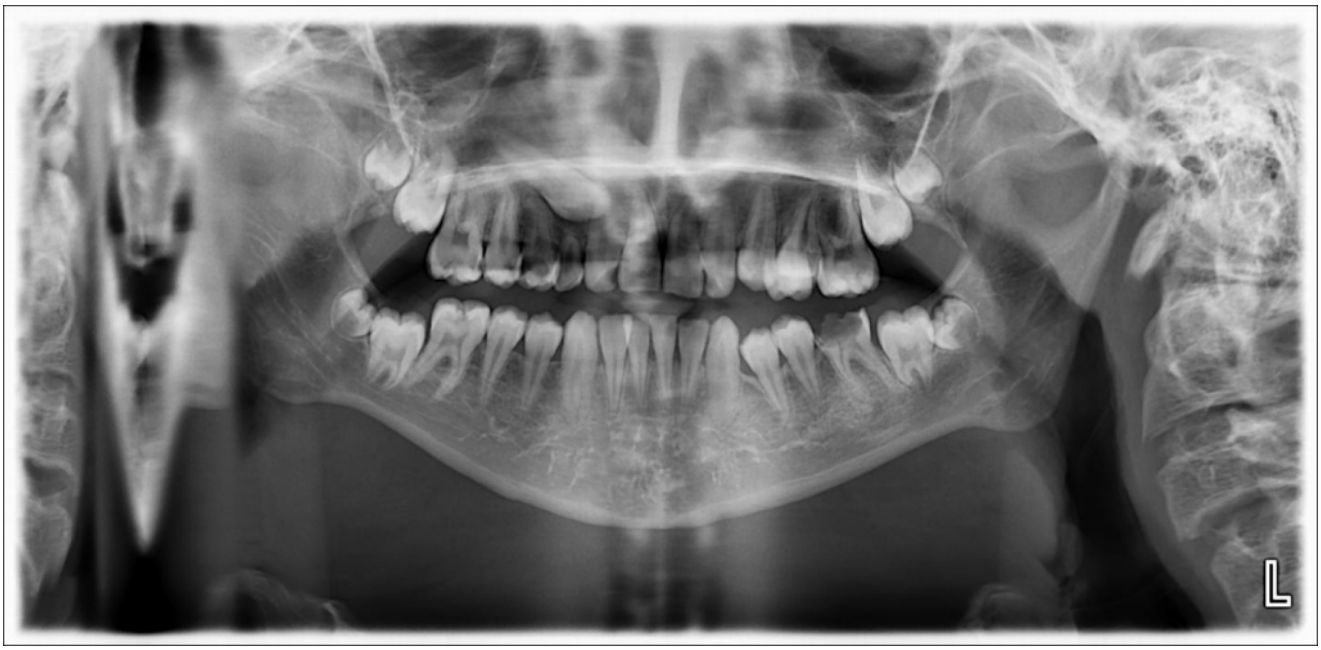


**Figure 2**
The patients panoramic x-ray

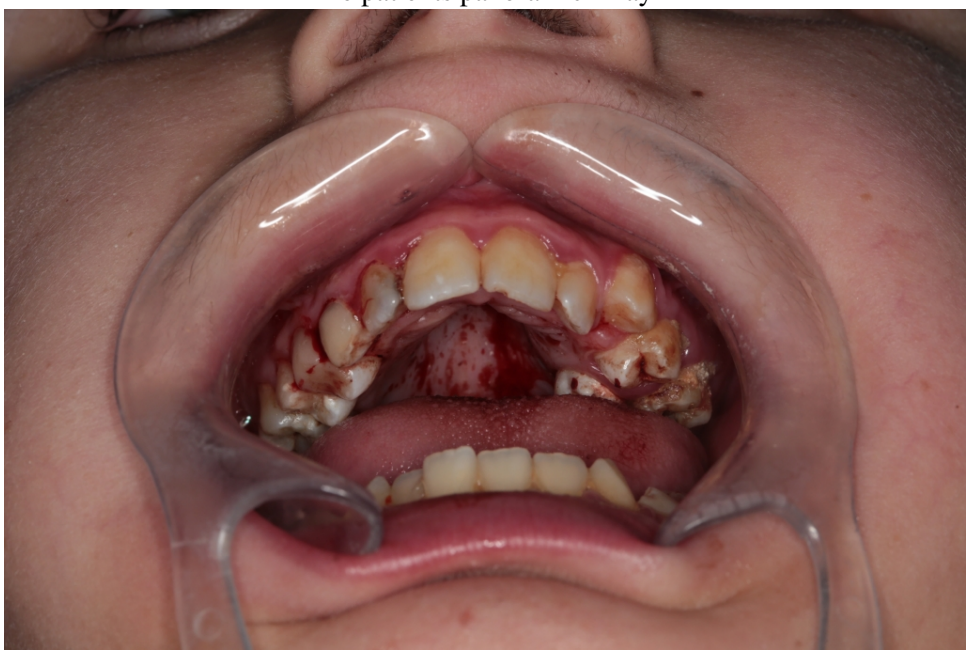

**Figure 3**
Intraoral image after the dental treatment

The process took a total of 75 minutes. Decurarization was performed via 2 mg kg-1 sugammadex administration at the end of the operation. For postoperative pain treatment, 10 mg $kg^{-1}$ paracatemol was infused for 30 minutes. Mr. ABU provided informed consent for publication of his case.

## Case Discussion

Although ES is a rare syndrome, it is the second most common chromosomal disorder accompanied by many anomalies. The rate of cardiac anomalies is 95% and it is the main responsibility of bad prognosis [1-3]. Our patient

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was being followed-up for pulmonary stenosis by cardiologists. İnfectious endocarditis prophylaxis is recommended in these patients before teeth extraction [4]. Dental treatments are performed under sedation or general anesthesia since mental retardation patients cannot cooperate. We preferred general anesthesia for our patient because the duration of the procedure was long. Patients usually require premedication before surgery [1].

Because the vein access at the beginning was not achieved after oral premedication we were induction with inhalation in the surgery room. For the cases where fatal anomalies are common in the literature, we recommend the shortest possible anesthesia and the least invasive procedure, so we preferred sevoflurane and propofol for our short duration of action.

We could not achieve vein access at the beginning, but after oral premedication, an IV line was secured and anesthesia was indused by inhalation in the surgery room. In the literature, the shortest possible anesthesia and the least invasive procedures are recommeded for patients with fatal anomalies so we preferred sevoflurane and propofol for our patient [1,5]. The article published by Sato et al., also mentioned a case of serious airway problem treated with sevoflurane [3,5]. Depolarising agents for muscle relaxant use are not recommended because of the risk of malignant hyperthermia, but short-acting nondepolarizans can be used [1,2]. The reason we prefer rocuronium bromide (*Myocron, vem ilaç sanayi*) as a muscle relaxant is that it has a specific antidote which is sugamadex(Bridion MSD ilaç). This drug can convert the rocuronium bromide connected block to any level.

We did not use atropine-neostigmine because of cardiac side effects. Alternative airway devices, such as laryngeal mask airway (LMA), must be available for the difficult anatomy of patients, especially for patients with facial and oral teeth anomalies [1]. In our patient, all necessary equipment and LMA for intubation were available and the intubation was successfully performed with videolaringoscopy. Assessing pain in these patients is difficult because of communication problems. We did not need any long lasting narcotic analgesics during the procedure.

We provided pain control with infiltration anesthesia. There are publications suggesting sublimation of narcotic analgesics [2,3], but we preferred remifentanil as a short-acting narcotics and as soon as possible effective narcotics.

After a while, our patient became agitated. We used paracatemol in the treatment of pain control and the patient calmed down. Different genetic variations related to the chromosome 18 have been mentioned in the literatüre [6,7]. In our case, multiduplication was detected in the short arm of the 18th chromosome. Genetic variations can affect the life span of the patient. Major cardiac anomalies may cause saturation level increase during positive pressure ventilation due to right-sided bag during general anesthesia. There are publications suggesting norepinephrine or dopamine for the treatment [8]. We have tried to protect the spontaneous breathing of the patient as much as possible and have kept the positive pressure ventilation short .

ES may be missed in many cases due to major cardiac anomalies at an early age. New treatment techniques developed every day, use of new drugs and increase in prenatal diagnosis by the use of genetic tests prolonged life expectancy in these patients. There are few reports in the literature that fully explain anesthesia management in these patients. Keeping the procedure as short as possible and avoiding any invasive procedures and complications, protect the patient from arrhythmia and cardiac and pulmonary hemostasis and provide hemodynamic stability and a smooth anesthetic process . By maintaining a strict adherence to the above procedures, patients with ES can be discharged in a healthy manner with rapid recovery at the end of a procedure.

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