Case Report/*Olgu Sunumu*

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# Difficult Intubation and General Anesthesia Management in a Pediatric Patient with Xeroderma Pigmentosum Requiring Emergency Laparotomy

## Acil Laparotomi Gereken Xeroderma Pigmentosumlu Pediatrik Hastada Zor Entübasyon ve Genel Anestezi Yönetimi

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**ABSTRACT**

Xeroderma pigmentosum (XP) is one of the rare neurocutaneous syndromes. The lesions occurring due to an impairment in the DNA repair mechanisms caused by ultraviolet light often require surgical interventions. There is no standardized general anesthesia procedure to be used in these patients. Besides, it is not clear whether sevoflurane leads to deterioration in the neurological condition. There are also other problems that may be encountered during difficult intubation and extubation. This may be the first reported pediatric patient with XP that underwent emergency laparotomy under general anesthesia with the successful use of sevoflurane, videolaryngoscopy, and sugammadex.

**Keywords:** Xeroderma pigmentosum, sevoflurane, airway management, sugammadex, laryngoscopy

**ÖZ**

Xeroderma pigmentosum (XP) nadir görülen nörokutanöz sendromlardandır. Genellikle ultraviyolenin yol açtığı DNA hasarını onaran sistemdeki bozukluk sonucu oluşan lezyonlar nedeniyle cerrahi gereksinimleri olmaktadır. Bu hastalarda uygulanacak genel anestezi konusunda bir standardizasyon geliştirilememiştir. Sevofluranın nörolojik durumda kötüleşme yapıp yapmadığı konusunda netlik yoktur. Zor entübasyon ve ekstübasyonda yaşanabilecek sıkıntılar da mevcuttur. Genel anestezi altında acil laparotomi geçiren XP'li pediatrik hasta sevofluran, videolaringoskopi ve sugammadeksin başarılı kullanıldığını bildirilen ilk vaka raporu olduğunu düşünmekteyiz.

**Anahtar sözcükler:** Xeroderma pigmentosum, sevofluran, hava yolu yönetimi, sugammadeks, laringoskopi

## INTRODUCTION

Among the neurocutaneous syndromes, Xeroderma pigmentosum (XP) is a rare disorder exhibiting an autosomal recessive inheritance pattern (1,2). The disease is caused by a defect in the nucleotide excision repair and the translation synthesis activity involved in the repair of DNA damage caused by an exposure to ultraviolet (UV) light (1). The disorder was described for the first time in the 1870s, and a total of 830 patients from 41 countries were reported until 1982. The United States (29%) and Japan (11%) have been the countries with the highest number of reported cases (2). According to the recent data, the incidence of XP is one patient per 22,000 people in Japan and one patient per 1,000,000 people in the Western countries (1). The disease symptoms include abnormal sunburn reaction, abnormal pigment change, cutaneous and non-cutaneous neoplasms, ocular abnormalities (photophobia, conjunctivitis, corneal drying and ulcer, keratitis, ocular neoplasms), neurologic abnormalities (microcephaly, low intelligence, abnormal speech, peripheral neuropathy, tremor, myoclonus, hearing loss, vocal cord paralysis, larynx dystonia), and immune abnormalities (1,2).

The lesions occurring in the face, head, and neck region due to UV light exposure in the patients with XP often require a surgical intervention. However, there are no guidelines in the literature regarding the anesthesia method to be used in such operations which will require general anesthesia. The most commonly reported challenges during anesthesia in these

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patients include difficulty with intravenous cannulations, ventilation, intubation and extubation, the risk of worsening neurological symptoms due to the use of inhalation anesthetics, and hypersensitivity to drugs (3). However, some studies reported no untoward effects of sevoflurane use in case series with XP (4).

The current report aims to contribute to the literature by presenting a patient that may be the first reported pediatric case of XP requiring emergency laparotomy due to an acute abdomen with successful use of sevoflurane in general anesthesia, the use of videolaryngoscopy in difficult intubation, and the use of sugammadex in extubation.

**Figure 1.** Obstruction image of the abdominal X-ray.

## CASE REPORT

A 17-year-old male patient (body weight 50 kg, height 163 cm) presented to our hospital with complaints of abdominal pain for the last three days and the inability to pass stool. The patient had a history of appendicitis at one year of age. Abdominal X-ray revealed air-fluid levels and the signs of obstruction (Figure 1). A laparotomy was planned with a pre-diagnosis of ileus. On preoperative assessment, the patient was realized to have XP, and pigmented lesions were observed in the face, arms, and legs. The patient also had microcephalus, cataract in both eyes, hearing impairment, facial skin tension, limited mouth opening (<3 cm), large upper teeth; and the patient's Mallampati score was 3 (Figure 2A). Head extension was normal. Laboratory tests revealed hyponatremia (132 mmol $L^{-1}$) and elevated CRP levels (20.78 mg $dL^{-1}$).

The patient was moved to the operating room, ECG was performed, and the patient underwent noninvasive blood pressure and $SpO_2$ monitorization. The heart rate was 107 bpm, the blood pressure was 135-69 mmHg, and $SpO_2$ was 99%. ST depression and negative T waves were observed on ECG. The patient came from the service with a 22 G peripheral venous line inserted, and he was started on intravenous 0.9% sodium chloride infusion. He was preoxygenated with 100% $O_2$ for 3 minutes then administered fentanyl 2 µg $kg^{-1}$, propofol 4 mg $kg^{-1}$, and rocuronium 1 mg $kg^{-1}$ intravenously to induce anesthesia. The patient with Cormack Lehane Score 3 was intubated using a 7.5 endotracheal tube (ID 7.5 mm) with a videolaryngoscope (Figure 2B). The ventilation parameters were adjusted to a 450 mL tidal volume, respiratory rate of 12 $min^{-1}$, PEEP of 4 $cmH_2O$, and end-tidal $CO_2$ of 35-40 mmHg in a volume-controlled mode. The anesthesia was maintained by 50% $O_2$, 50% dry air, and 1-2% sevoflurane. Dense adhesions

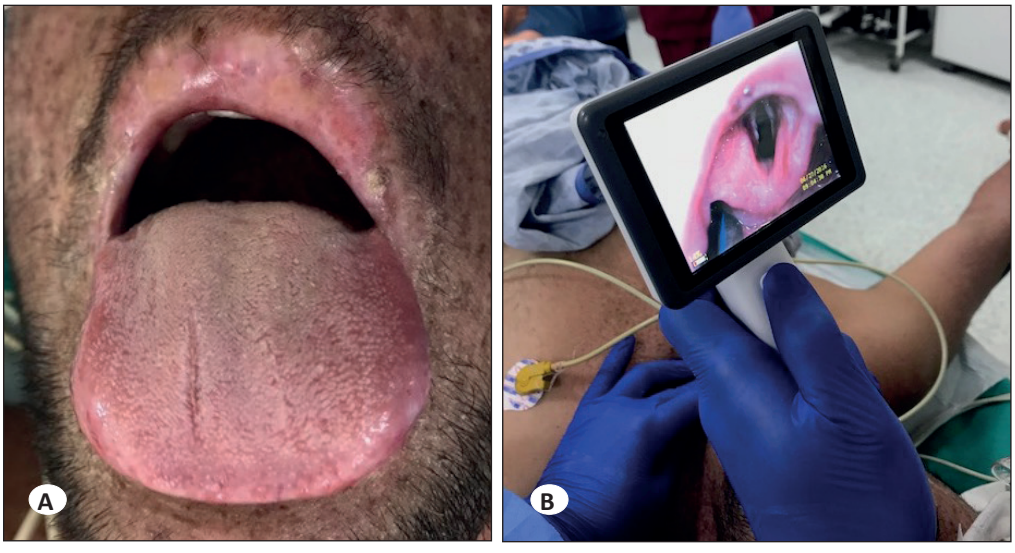


**Figure 2.** Mallampati score III **(A)**, and video laryngoscopic image **(B)** in a patient with Xeroderma pigmentosum.

were discovered among the bowel loops, and adhesiolysis was performed. The patient's respiratory efforts resumed after general anesthesia that lasted 120 minutes, and the patient was extubated with the administration of sugammadex 4 mg $kg^{-1}$ without any problem. No complications occurred in the postoperative period, and the patient was discharged after five days.

## DISCUSSION

The patients with XP require multiple surgical interventions due to skin lesions, eye lesions, and malignancies that arise from their skin's abnormal reaction to UV light (3). Although rare, these patients may require more extensive and major surgeries. The patient presented in the current case report is the first pediatric patient with XP undergoing surgery due to acute abdomen.

Skin lesions such as hyperpigmentation, actinic keratosis, irregular poikiloderma, telangiectasia, and cutaneous neoplasms in patients with XP may complicate establishing vascular access (4,5). Inhalation anesthesia may be required if the attempts to establish a vascular line fail in a pediatric patient. However, difficulties may be encountered in the mask ventilation and the intubation of patients with XP due to the presence of cutaneous nodular neoplasms in the face, large tongue, incisor teeth protrusions, limited mouth opening, and high Mallampati scores (4,5). As it was performed in the present patient, the use of a videolaryngoscope for endotracheal intubation may help safely secure the airway. The reason why our first choice is videolaryngoscope due to both the difficult intubation criteria of the patient (facial skin tension, limited mouth opening <3 cm, large upper teeth, Mallampati score 3) and our clinical approach to all patients who were taken to emergency surgery because of the COVID-19 pandemic.

Inhalation agents and drugs used in general anesthesia have different effects over this particular patient group. Isoflurane and sevoflurane are known to be associated with a deterioration in neurological symptoms, and it is stated that these patients have increased hypersensitivity against benzodiazepines, opioids, and muscle relaxants (3). However, no study in the literature has suggested a neurological impairment in pediatric patients receiving sevoflurane. Total intravenous anesthesia (TIVA) has been preferred in some pediatric cases to avoid inhalation anesthetics. It is stated that BIS (Biscpectral index) monitorization has also been performed in patients receiving TIVA (6,7). It is an outstanding finding that short-acting muscle relaxants are used in reduced doses. The monitorization of neuromuscular blockade may be useful in avoiding prolonged drug effects (7). Some studies have reported using an agent to reverse the neuromuscular blockade without specifying the reversal agents (5). In the present study, sugammadex was used to avoid the prolonged effects of rocuronium and facilitate extubation. Therefore, the present patient is the first pediatric patient with XP in the literature in whom sugammadex was used. Anesthesia managements applied to pediatric patients with XP in the last 20 years are shown in Table I.

Another consideration with these patients is the UV radiation sources in the operating room. The surgical spotlights, headlamps and laparoscopic light source emitted significant radiation; therefore, they must be covered with UV-filtering material (19). Since we could not provide such a material for our patient who was operated under emergency conditions, we had to operate him in an operating room with LED lights that do not emit UV radiation, and with an open surgical method.

There is no standardized protocol for administering general anesthesia in the patients with XP. In addition, the effects of inhalation anesthetics and drugs used in general anesthesia are unknown. The authors of this article consider that sevoflurane can be utilized in pediatric patients with XP who do not have established vascular access, that using a videolaryngoscope can facilitate intubation in these patients who frequently have a difficult airway, and that using muscle relaxants whose effects are reversed by sugammadex can be beneficial.

## AUTHOR CONTRIBUTIONS

**Conception or design of the work:** CSO, KES

**Data collection:** CSO

**Data analysis and interpretation:** CSO, KES

**Drafting the article:** CSO

**Critical revision of the article:** KES

All authors (CSO, KES) reviewed the results and approved the final version of the manuscript.

**Conflict of Interest:** The authors have no conflicts of interest to declare.

**Informed Consent:** Written informed consent was obtained from the guardian (parents) of patient who participated in this case.

**Table I.** Summary of Reported Anesthetic Managements in a Pediatric Cases of Xeroderma Pigmentosum

| Case | Patient years/Sex | Operation | Intubation | Induction | Neuromuscular blocker | Maintenance of anesthesia |
|---|---|---|---|---|---|---|
| Sethi et al. 2019 (5) | 10 / F | Eye nodule excision | LMA (IGel) | Propofol, Ketamine | Atracurium | Dexmedetomidine Nitrous oxide |
| Karami et al. 2019 (10) | 6 / M | Skin tumors excisional biyopsi | LMA | Propofol | None | TIVA (propofol, remifentanil) Nitrous oxide |
| Zarrouki et al. 2017 (4) | Case series | Plastic surgery | Laryngoscopy | Sevoflurane | Rocuronium | Sevoflurane |
| Parajuli et al. 2016 (3) | 12 / M | Enucleation of eyeball | Laryngoscopy | Propofol, Dexmedetomidine | None | TIVA (propofol, dexmedetomidine) |
| Bharti et al. 2016 (11) | 8 / M | Scalp excision and grafting | Laryngoscopy | Sevoflurane | Atracurium | Dexmedetomidine |
| Shah et al. 2015 (7) | 12 / F | Basal cell neoplasm excision | Laryngoscopy | Propofol, Fentanyl | Succinylcholine | TIVA (propofol) |
| Ubale et al. 2015 (12) | 6 / M | Dental extraction | Nasal intubation | Propofol | Succinylcholine | TIVA (propofol) Nitrous oxide |
| Goyal et al. 2015 (13) | 16 / F | Scalp excision | None | Dexmedetomidine Ketamine | None | Dexmedetomidine Ketamine |
| Mahin et al. 2014 (14) | 9 / M | Parotid abscess drainage | LMA | Propofol | None | TIVA (propofol, remifentanil) Nitrous oxide |
| Mulimani et al. 2013 (15) | 9 / F | Excision of basal cell carcinoma | Laryngoscopy | Propofol | Succinylcholine Atracurium | Nitrous oxide |
| Gupta 2011 (16) | 8 / M | Eye lesion excision | Laryngoscopy | Propofol | Succinylcholine | TIVA (propofol) Nitrous oxide |
| Soen et al. 2006 (17) | 15 / F | Clubfoot | ? | ? | ? | TIVA (propofol, fentanyl) |
| Brunner et al. 2004 (8) | 6 / F | Skin covering the nose resection | Laryngoscopy | Sevoflurane Fentanyl | Mivacurium | Sevoflurane |
| Oliveira et al. 2003 (9) | 7 / F | Ocular lesion excision | Laryngoscopy | Sevoflurane Propofol | Succinylcholine | Sevoflurane |
| Masuda et al. 2002 (18) | 17/ F | Tracheostomy | ? | ? | ? | TIVA (propofol, fentanyl) |

**F:** Female, **M:** Male, **LMA:** Laryngeal mask airway, **TIVA:** Total intravenous anesthesia.

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