

## Anesthetic Management of a Patient with Pompe Disease: Our Experiment

### Pompe Tanılı Bir Olguda Anestezi Uygulamamız

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#### ABSTRACT

Pompe disease is a fatal autosomal recessive glycogen storage disease presenting with hypertrophic cardiomyopathy, widespread myopathy and hypotonia. In this presentation, we aimed to present our anesthetic approach to 12 teeth extraction planned non-cooperative, 34-month-old patient who were being followed-up in an external center with Infantile Pompe Disease.

**Key Words:** Pompe disease, sedoanalgesia, monitorized anesthesia care

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#### ÖZET

Pompe hastalığı, hipertrofik kardiyomyopati, hipotoni ve kas güçsüzlüğü ile seyreden otozomal resesif geçişli fatal bir glikojen depo hastalığıdır. Bu yazıda infantil pompe hastalığı tanısı ile dış merkezde takipli, 12 diş çekimi planlanan ancak kooperasyon kurulamadığından tarafımıza danışılan 34 aylık olguya anestezi yaklaşımımızı sunmak istedik.

**Anahtar Sözcükler:** Pompe hastalığı, sedoanaljezi, monitörize anestezi bakımı

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#### INTRODUCTION

Pompe disease is an autosomal recessive storage disease in which glycogen accumulates in tissues without hydrolysis due to the lack of lysosomal alpha-glycosidase (acid maltase) enzyme. There are three types, infantile, with decreased enzyme activity, juvenile with absent enzyme activity and adult type. Infantile type usually results with death in the first two years due to hypertrophic cardiomyopathy, widespread myopathy, hypotonia, growth retardation and respiratory failure (1-4).

In this presentation, we aimed to present our anesthetic approach to 12 teeth extraction planned non-cooperative, 34-month-old patient who were being followed-up in an external center with Infantile Pompe Disease, who had hypertrophic cardiomyopathy, hypotonia, mental-motor retardation.

#### CASE REPORT

A 14-kg 34-month-old female patient with infantile Pompe disease who received enzyme treatment with 15-day periods was consulted because of lack of cooperation during tooth extraction. In preoperative evaluation; The patient was diagnosed as having Mallampatia 3, with mouth opening reduced, macroglossia, wheezing, hepatosplenomegaly, hypotonia, she was planned to appropriate fasting period under appropriate anesthesia care (MAB). The patient was moved to operation room without premedication, EMLA® administered to left hand dorsum, monitorized by non-invasive means, peripheral Intravenous (IV) cannulation performed while nasal oxygen administered from 4lt/min.

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Infiltration anesthesia was performed with a mixture of Artikain HCl-Epinefrin HCl (Ultrakain®) + Lidocaine HCl (Aritmal®) when Ramsey Sedation Scale (RSS) was detected to be 4, after IV application of 1.5 mg kg-1 ketamine (Ketal®). During the 15 min procedure, 0.5 mg kg-1 IV ketamine was administered to the patient as additional dose, at the end of the procedure patient was followed up for 10 minutes and moved to ward as hemodynamically stable, with RSS: 1.

## DISCUSSION

Monitored anesthesia care (MAC) is defined as to monitor the patient by providing additional medication which anxiolytic, amnesic, analgesic and hypnotic to diagnose or to treat the patient for therapeutic intervention. The main purpose of MAC is ensuring patient comfort, removal of anxiety, pain control, hemodynamic stability, immobilization of patient (5). With the combination of ketamine and infiltrational anesthetics, which we prefer because of the possibility of malignant hyperthermia and presence of hypertrophic cardiomyopathy. With ketamine's analgesic, hypnotic and amnesic effects, it doesn't suppress pharyngeal and laryngeal reflexes, does not cause cardiovascular and respiratory depression (3,4, 6) We believe that we have performed the procedure without side effects, without the need for additional anesthetic stabilization.

## Conflict of interest

No conflict of interest was declared by the authors.

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