

# Repeated General Anesthesia in a Patient With Noonan Syndrome

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Noonan syndrome (NS) is an autosomal dominant disorder characterized by facial anomalies, short stature, chest deformity, congenital heart diseases, and other comorbidities. The challenges faced during anesthetic management of patients with NS could be due to congenital heart diseases, hemostatic disorders, and airway anomalies. Here we describe dental treatment under general anesthesia performed for a 28-year-old man with NS. He had characteristic features of NS along with mild pulmonary valve stenosis. Dental treatment under general anesthesia was performed successfully on 13 occasions with nasotracheal intubation under curve-tipped suction catheter guidance or insertion of a reinforced laryngeal mask airway. This case suggests that for patients with NS, who might present several challenges, dental anesthesiologists should consider the extent of the patient's disorders to enable them to perform dental treatment safely under general anesthesia.

**Key Words:** Repeated general anesthesia; Noonan syndrome.

Noonan syndrome (NS) is an autosomal dominant disorder and was first reported by Noonan and Ehmke.<sup>1</sup> The primary features of this multisystem disorder include hypertelorism, low-set ears, down-slanting eyes, a webbed neck, congenital heart diseases, short stature, chest deformity, and intellectual impairment.<sup>2,3</sup> NS affects males and females equally and has an estimated incidence of 1 in 1000 to 1 in 2500 live births.<sup>2,4</sup>

The challenges faced during anesthetic management of patients with NS could be due to congenital heart diseases, hemostatic disorders, and airway anomalies.<sup>5-8</sup> The present report describes repeated administration of general anesthesia performed by adopting nasotracheal intubation or reinforced laryngeal mask airway insertion for a patient with NS at the time of dental treatment in our hospital.

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## CASE HISTORY

Dental treatment under general anesthesia was scheduled for a 28-year-old man with NS (height, 145 cm; weight, 23 kg). In November 2011, he had visited our hospital with his mother with the complaint of several carious teeth. His mother informed that he had been diagnosed with NS in a general hospital near their residence when he was 1 year old. He underwent medical care for pulmonary valve stenosis, scoliosis, bone fragility, epilepsy, and intellectual impairment at the general hospital along with the prescribed follow-up. In March 2011, an oral surgeon at the hospital extracted his third molars under general anesthesia. Although he had received dental treatment in a dental office near his residence in his early childhood under physical restraint, he began to refuse dental treatment as he got older. His mother asked us to provide dental treatment for him.

The patient presented characteristic features of NS, including hypertelorism and low-set ears, in addition to the conditions mentioned above. A mild peak pressure

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gradient of the pulmonary valve was reported and needed no treatment. He used a wheelchair and was unable to communicate with us. Although we could not inspect his mouth well, we estimated that he had 10 carious teeth and an approximately 35-mm mouth opening. Thereafter, we planned to perform dental treatment under general anesthesia because he would not open his mouth on request.

On day 1 of the dental treatment, the patient was placed on a dental chair in the supine position and intravenous catheterization was performed. He was then given 5% sevoflurane mixed with 6 L/min of nitrous oxide gas and 3 L/min of oxygen through a face mask, with soft cushions placed under his left shoulder and his knees to reduce the burden on his excessively curved spine and legs. Lubricant (Caine Zero Jelly; FUJIFILM Medical Co Ltd) mixed with 1% lidocaine containing 1 : 100,000 adrenaline was applied to both nostrils to improve slippage of the nasotracheal tube and prevent nasal bleeding. Subsequently, nasal endotracheal intubation was attempted gently under general anesthesia maintained with 20 mg of rocuronium, 20 mg of propofol, 0.3 mg of atropine sulfate, and continuous infusion of propofol at 4.3 mg/kg/h along with remifentanyl hydrochloride at 0.18  $\mu$ g/kg/min. At the first attempt to insert an endotracheal tube (Parker Flex-Tip, Preformed Nasal Cuffed; Parker Medical) into the right nostril, the dental anesthesiologist was unable to advance the tube past the posterior area of the nasal cavity. After withdrawing the tube from the nostril, the dental anesthesiologist inserted a suction catheter through the tube with the catheter tip emerging from the distal end of the tube.<sup>9</sup> He again inserted the endotracheal tube with the catheter into the nostril and pulled out the distal end of the catheter from the mouth using Magill forceps. Subsequently, he succeeded in advancing them into the pharynx by pulling up the distal end of the catheter and the tube tip. After removing the catheter, he achieved nasal intubation smoothly under direct laryngoscopy. Nasal bleeding was not observed. Anesthesia was then maintained with a continuous infusion of propofol at 4.3–7.0 mg/kg/h and remifentanyl at 0.07–0.18  $\mu$ g/kg/min, while the dentist in charge scaled all teeth and performed metal core cavity preparations on 2 carious teeth. The patient left the hospital after an uneventful period of 3 hours after extubation.

In total, the patient underwent dental treatment under general anesthesia on 13 occasions at our hospital. On 11 of these occasions, general anesthesia was administered with nasal endotracheal intubation, using a similar method each time. On the remaining 2 occasions we used a flexible reinforced laryngeal mask airway because the dentist in charge did not require nasal endotracheal

intubation for the dental procedures. The same agents were used to introduce and maintain anesthesia, except for rocuronium. We advanced the laryngeal mask airways after straightening the tips of the airways that were bent behind and caught at the posterior wall of the pharynx, thus succeeding in placing them in acceptable positions.

We observed a tendency of bleeding after pulpectomy of the left first and second molars of the mandible on the fourth occasion when the patient attended our hospital for treatment. Although the dentist in charge extracted the left second molar of the maxilla after preparation of a surgical splint, this bleeding tendency was not observed in the patient's last treatment under general anesthesia.

## DISCUSSION

We successfully performed nasotracheal intubation and insertion of a reinforced laryngeal mask airway repeatedly in a patient with NS. Several studies have reported difficult intubation in cases of NS because of limited mouth opening, webbed neck with cervical spine anomalies, and other craniofacial anomalies; in such cases, intubation was achieved with fiber-optic bronchoscopy or awake direct laryngoscopy.<sup>5–8</sup> Our patient might have had some deformities in the posterior area of the posterior nasal and oral cavities, because advancement of the nasotracheal tubes and reinforced laryngeal mask airways was challenging in these areas. We had intended to utilize other techniques such as oral endotracheal intubation and insertion of a flexible reinforced laryngeal mask airway on an occasion when we failed to pass an endotracheal tube through the nasal cavity in the way described above.

Most patients with NS have pulmonary valve stenosis, and a smaller number have other heart diseases such as hypertrophic obstructive cardiomyopathy, atrial septal defect, ventricular septal defect, and patent ductus arteriosus.<sup>2,4</sup> Moreover, patients with NS may have hemostatic disorders as well.<sup>2,4</sup> These factors affect decisions regarding anesthetic management of such patients.<sup>6</sup> The present patient had mild pulmonary valve stenosis but no other congenital heart disease; consequently, we did not consider this to be significant in the anesthetic management. Although results of the hemostatic function tests were within normal limits, we advanced the nasotracheal tube gently using lubricant mixed with adrenaline and succeeded in preventing nasal bleeding, because cases of postoperative bleeding have occurred despite normal *in vitro* clotting assays and platelet counts.<sup>10</sup> However, as the bleeding tendency

observed on pulpectomy was unpredictable but bleeding was possible, we prepared a hemostatic splint before tooth extraction.

We had to secure an intravenous route under inhalation anesthesia and prepare suitable cushions to reduce external forces from restrainers and dental chairs.

Because patients with NS have several challenges, as described above, dental anesthesiologists must consider the extent of these disorders to enable them in performing dental treatment safely under general anesthesia.

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