

Successful Airway Management In A Patient With Progeria

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Citation

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Abstract

We present the case of a twenty one year old female with progeria for dental surgery, in whom tracheal intubation was extremely difficult secondary to multiple anatomic factors. These included poor mouth opening, small mandible and neck stiffness. The trachea was secured by using a flexible fiberoptic bronchoscope. Practitioners caring for patients with progeria should be aware that a intubation of the trachea may be difficult or impossible using direct laryngoscopy, and prepare for an alternative plan.

INTRODUCTION

Progeria is a genetic disorder that resembles a state of accelerated aging. The majority of those afflicted have Hutchinson-Gilford progeria syndrome, which is usually caused by a single nucleotide defect in the LMNA gene which codes for the lamin A and C proteins and is probably always a spontaneous mutation [1, 2]. Although babies with progeria appear normal at birth, by one year of age marked growth retardation usually becomes evident. These children go on to suffer from baldness, tissue atrophy, osteoporosis, atherosclerosis, hypertension and other afflictions normally associated with the elderly; intelligence is unaffected. Death generally occurs in the teen years from cardiovascular illness. The stunted development also affects facial structures such as the mandible, maxilla and oral aperture, leading to small facies and dental crowding [3, 4]. Given these facial features one should predict that these patients would be difficult to intubate. We report a patient with Progeria and a difficult airway.

CASE REPORT

A 21-year-old woman with progeria presented to Rady Children's Hospital in San Diego for dental surgery (figure 1). She needed to have several teeth extracted, but because of her inability to adequately open her mouth in the dentist's office she was scheduled to have the procedure done in the operating room under general anesthesia.

The patient had been diagnosed with progeria at a young age but was doing fairly well at the time of surgery. She had hypertension, but no other significant cardiovascular disease. Her daily medications were atorvastatin and metoprolol. She

had had an orthopedic surgical procedure at age 19, and her intubation at that time had reportedly taken two hours and required a fiberoptic bronchoscope.

On examination, she bore the typical stigmata of progeria: a shrunken, bird-like face, a relatively large cranium, partial baldness, wrinkled and scaly skin, poor joint mobility, short stature, micrognathia, and crowded teeth (Figure 1). She had a Class Four Mallampati score, three centimeters of hyomental distance, and a severely limited range of neck motion. She could only open her mouth two centimeters and could barely stick out her tongue.

Figure 1



The patient adamantly refused an awake intubation, but did consent to a pre-operative IV. In the operating room the IV and routine monitors were placed and then the patient was induced via mask with 65% N₂O and sevoflurane. After an easy mask airway was assured, propofol 80 mg and mivacurium 5 mg were administered intravenously. With great difficulty, a small intubating oral airway was inserted into the oropharynx and the tongue was gently displaced with a triangular clamp over gauze. A second anesthesiologist assisted with displacing the jaw and opening

the mouth. The trachea was entered under direct vision with the bronchoscope and a 6.0 endotracheal tube was advanced; its position was adjusted to 2 cm above the carina using the fiberoptic scope. The tube was secured, the surgery proceeded, and the patient was extubated without difficulty when she was awakened after the case.

DISCUSSION

Patients with progeria who require anesthesia can present multiple challenges [5, 6]. These include premature end organ disease, hypertension, difficult intravenous access, and a potentially difficult airway. However, a patient with progeria should never present as a “surprise” difficult airway. There are several examples in the literature of the intubation challenges that they can present [3, 4, 5], and the combination of micrognathia, dental crowding, small jaw size and unsupple neck that typifies their airway exam should raise red flags. A controlled awake or asleep fiberoptic intubation is a judicious option when an endotracheal airway is required. Even though our patient was of adult age, she presented the same challenge as some of our pediatric patients in her inability to fully cooperate with an awake intubation. This is not surprising in a young patient with chronic disease, but because of this we opted to do an inhalational induction with spontaneous respiration and assure that the mask airway was adequate before administering propofol and mivacurium to optimize intubating conditions. Had she been a difficult mask, we

might have opted to awaken her instead of proceeding. Due to their small nares, nasal intubation is impractical in patients with progeria, so an oral route was chosen for this case and the surgeon agreed to work around the endotracheal tube. Laryngoscopy would likely not have been possible because of the inability to open this patient’s mouth more than two centimeters. It was helpful to have a second anesthesia provider present from the start of the case to offer experienced assistance during the induction. Our selection of a planned asleep oral fiberoptic approach allowed the intubation to be quick and uncomplicated and was a simple and safe choice for this patient.

References

1. Sarkar PK, Shinton RA. Hutchinson-Guilford progeria syndrome. *Postgrad Med J*. 2001; 77:312-17
2. Kudlow BA, Kennedy BK. Aging: Progeria and the Lamin Connection. *Curr Biol*. 2006 Aug 22;16(16):R652-4.
3. Batstone MD, Macleod AW. Oral and maxillofacial surgical considerations for a case of Hutchinson-Gilford progeria. *Int J Paediatr Dent*. 2002 Nov;12(6):429-32.
4. Arai T, Yamashita M. An abnormal dentition in progeria. *Paediatr Anaesth*. 2002 Mar;12(3):287.
5. Liessmann CD. Anaesthesia in a child with Hutchinson-Gilford progeria. *Paediatr Anaesth*. 2001;11(5):611-4.
6. Nguyen NH, Mayhew JF. Anaesthesia for a child with progeria. *Paediatr Anaesth*. 2001 May;11(3):370-1.

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