Successful Airway Management In A Patient With Progeria
S Merritt, M Greenberg

Citation

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.

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
Successful Airway Management In A Patient With Progeria

the mouth. The trachea was entered under direct vision with
the bronchoscope and a 6.0 endotracheal tube was advanced;
it's 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
3. Batstone MD, Macleod AW. Oral and maxillofacial surgical considerations for a case of
5. Liessmann CD. Anaesthesia in a child with Hutchinson-Gildford progeria.
6. Nguyen NH, Mayhew JF. Anaesthesia for a child with progeria.
Author Information

Sidney Merritt, MD
Associate Clinical Professor of Anesthesiology, University of California, San Diego Medical Center

Mark Greenberg, MD
Clinical Professor of Anesthesiology and Pediatrics, University of California, San Diego Medical Center