

## Cherubism with Difficult Laryngoscopy and Tracheal Intubation

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Cherubism is a rare familial disease causing enlargement of the mandible and sometimes the maxilla that is often associated with a difficult endotracheal intubation. We describe such a case in which marked difficulty with laryngoscopy and endotracheal intubation was encountered.

## REPORT OF A CASE

The patient initially was evaluated for mandibular enlargement at 2 yr of age. She had a negative family history for cherubism, and her older brother, age 6 yr, was unaffected. Radiographs taken at the time demonstrated alveolar ridge abnormalities and mandibular enlargement consisting of bone cysts with hypertrophy. This mandibular enlargement was followed until age 8 yr, when she underwent a surgical procedure at another hospital for mandibular reduction to allow eruption of her permanent teeth. There was no history of any anesthesia-related problems. Prior to this time, she was evaluated for short stature and demonstrated normal growth hormone and thyroid function studies. At age 11 yr, she was evaluated for easy bruising and thrombocytopenia but demonstrated normal megakaryocytes on a normal bone marrow biopsy. Shortly thereafter, she was hospitalized to undergo mandibular reduction for cosmesis.

Preoperative evaluation on admission demonstrated a pleasant, small for stated age, white girl with remarkable mandibular enlargement (fig. 1). She demonstrated normal neck flexion and no trismus, despite the size of her mandible. The remainder of her physical examination was unremarkable for the present discussion. Figure 2 is a radiograph of her mandible demonstrating the cystic bony enlargement. Her preoperative laboratory studies were significant for thrombocytopenia of  $85,000/\text{mm}^3$  but a normal bleeding time. She was taken to the operating room, and general anesthesia was induced with halothane,  $\text{N}_2\text{O}$  and  $\text{O}_2$ . The  $\text{N}_2\text{O}$  was discontinued, and then multiple attempts at endotracheal intubation were undertaken. These included oral direct laryngoscopy, nasotracheal intubation with and without direct laryngoscopy, pediatric fiberoptic bronchoscopy, and retrograde insertion of a 17-gauge epidural catheter through the cryothyroid membrane. All efforts at intubation proved unsuccessful. At no time were the laryngeal structures identified during laryngoscopy during which ventilation was always spontaneous. In between attempts at intubation, ventilation was assisted. During the last attempts at intubation, the patient demonstrated unifocal and multifocal ventricular premature contractions and had one episode of bradycardia (40 bpm). Because of these dysrhythmias and the lack of progress the case was cancelled. At that

point auscultation of the chest revealed rales and decreased breath sounds over the right upper lung field. It was felt the patient may have aspirated blood and oral secretions. There was no apparent regurgitation or vomiting. A chest roentgenograph revealed infiltration of both right upper and lower lobes, suggestive of aspiration. She was tachypneic at a rate of  $40 \text{ breaths} \cdot \text{min}^{-1}$ . She was treated with antibiotics. Overnight the respiratory rate returned to normal and auscultation of the chest was normal. A repeat chest roentgenograph the next morning demonstrated almost complete clearing, with a small amount of infiltrate in the right upper lobe. She discharged from the hospital and returned 3 months later for mandibular reduction. Her physical findings at this hospitalization were unchanged. She was taken to the operating room with plans for a tracheostomy to be performed if attempts at nasotracheal intubation were unsuccessful. Following iv induction of general anesthesia with 4 mg/kg thiopental and spontaneous assisted ventilation with halothane/ $\text{O}_2$ , 4% lidocaine was applied topically to the nasopharynx and oropharynx, and a blind nasotracheal intubation



FIG. 1. An 11-year-old girl with cherubism.

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with a stylet was performed atraumatically and rapidly. No attempt was made at direct laryngoscopy for visualization of cord structures. The patient underwent her operative procedure, which entailed a 2,100 ml blood loss, for which she received replacement with two units packed red blood cells, two units of whole blood, and six units of platelet concentrates. She had an unremarkable postoperative course in the surgical intensive care unit, and her trachea was extubated on the first postoperative day. She was discharged from the hospital on the eighth postoperative day.

#### DISCUSSION

The term "cherubism" was coined in 1933 by Jones<sup>1</sup> in his report of three siblings with painless mandibular enlargement. Cherubism is a disease of early childhood and consists of painless mandibular enlargement, with and without maxillary involvement, and progresses rapidly over the course of several years. The syndrome generally regresses during puberty and on into adult life. Adults affected in early childhood may demonstrate only mild mandibular enlargement. Maxillary regression generally precedes mandibular regression.<sup>2</sup> With maxillary enlargement and subsequent elevation of the floor of the orbit, patients may demonstrate increased exposure of the lower sclera, giving the look of "eyes raised to heaven,"<sup>2</sup> for which Jones coined the term cherubism. A number of synonyms have been used to report subsequent cases, including familial multilocular cystic disease, bilateral giant cell tumors, familial fibrous swellings, familial intraosseous fibrous swellings, familial fibrous dysplasia, familial osseous dysplasia, and hereditary fibrous dysplasia.<sup>2</sup> Cherubism demonstrates an autosomal dominant inheritance, with variable penetrance ranging from 80 to 100% in male patients to 50–70% in female patients.<sup>3,4</sup>

The diagnosis of cherubism must be made on clinical presentation, family history, physical examination, radiographic variables, and clinical follow-up.<sup>5</sup> The radiographic lesions of cherubism are multiple, well-defined, multilocular radiolucencies that may contain displaced or unerupted teeth that appear to "float in the radiolucent areas."<sup>6,7</sup>

The treatment of cherubism is generally conservative because the course usually proceeds toward spontaneous remission.<sup>2,8</sup> In those cases where intervention is deemed necessary, surgical curettage and reconstruction is the treatment of choice.<sup>5,8,9</sup> Radiation is not indicated due to long-term risks,<sup>2,5,8</sup> and steroids have not been shown to be beneficial.<sup>5</sup> Nasotracheal intubation is the preferred airway route for mandibular reduction because of the oral approach to the procedure.

The anatomic reason for the difficulty with intubation is the enlargement of the mandible. During intubation in normal individuals the area of soft tissue bounded on three sides by the mandible and inferiorly by the hyoid bone is displaced by the laryngoscope, and visualization



FIG. 2. Anterior-posterior radiograph of cherubism. Arrows indicate the inside of the ramus of the mandible. The space between the arrows is the potential displacement area. This is the same 11-year-old girl as Figure 1.

of the larynx is possible. In cherubism this potential displacement area is encroached upon by varying amounts of mandibular enlargement (fig. 2). In our patient, the mandibular rami were so close in the midline that there was no area to displace the tongue and soft tissue during laryngoscopy. Preoperative testing such as opening the mouth and extending the neck would not reveal the problem. The preoperative examination to determine the extent of the problem involves palpation of the soft tissue of the potential displacement area. Varying degrees of encroachment on this area lead to varying degrees of difficulty with laryngoscopy. Similar problems are found in the Pierre Robin syndrome, Goldenhar's syndrome, and in Treacher Collins syndrome.

Once the difficult airway is identified, then various therapeutic maneuvers to accomplish intubation must be considered and an anesthetic plan with various contingencies developed. Blind nasotracheal intubation with a stylet in the tube, a technique recently described, proved to be quite useful in this case and in similar situations.<sup>10</sup>

Awake intubation was discussed with the patient, and she refused the technique. Awake intubation was suggested because of the previous difficulty with intubation and apparent aspiration of blood and airway secretions. It was not felt that she aspirated gastric contents, and this was the reason for the rapid recovery. The other diagnostic possibility was negative pressure pulmonary edema secondary to airway obstruction. The persistence of the infiltrate was felt to be more typical of aspiration.

The objective of this case report is to highlight the unusual airway problem presented by this patient. Cherubism with its mandibular bony enlargement provided a resistance to a displacement of the tongue for visualization of the larynx on direct laryngoscopy and the resultant difficult intubation.

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## Seizure-like Movements during a Fentanyl Infusion with Absence of Seizure Activity in a Simultaneous EEG Recording

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Recent case reports of fentanyl-induced seizures in patients have generated much discussion and concern.<sup>1-5</sup> Opiates, including fentanyl in sufficiently high doses, can cause electroencephalographically proven seizures in experimental animals.<sup>6-8</sup> EEG-documented seizures due to fentanyl in humans have not been reported, including the above-cited case reports, which did not have the benefit of EEG recording during the event. We report a case of seizure-like muscle movements in a patient during a fentanyl infusion with simultaneous EEG recording and measurement of fentanyl plasma concentrations, arterial blood gas values, and hemodynamic values.

### REPORT OF A CASE

A 73-year-old, 81-kg man was scheduled for a right below-knee amputation for a chronic foot ulcer. He had peripheral vascular

disease, a 10-yr history of adult-onset diabetes mellitus, chronic atrial fibrillation, and a myocardial infarction 20 years previously. He reported no angina for many months, nor did he have symptoms of congestive heart failure, neurologic disease, or stroke. His medications were NPH insulin 40 units each morning, isosorbide dinitrate 10 mg po bid, and digoxin 0.125 mg qd. Except for the foot ulcer, the physical and neurologic examinations were unremarkable. Laboratory data were also normal.

The night before surgery he gave informed consent to participate in an on-going study of the EEG effects of narcotic infusions,<sup>9</sup> which included frequent blood sampling for narcotic concentrations. He was brought to the operating room without receiving premedication. EEG electrodes and leads for a 4-channel EEG were applied (FP<sub>1</sub>-O<sub>1</sub>, FP<sub>2</sub>-O<sub>2</sub>, Cz-O<sub>1</sub>, Cz-O<sub>2</sub>).<sup>10</sup> Arterial and venous catheters were inserted, and he was given pure oxygen to breathe via a face mask. A 5-min baseline EEG was recorded with the patient resting quietly with his eyes closed. During this period he received pancuronium 1 mg and glycopyrrolate 0.2 mg iv. Heart rate and mean arterial pressure were recorded every 30 s. Baseline heart rate was 95 bpm, and mean arterial pressure was 92 mmHg. After the baseline period an infusion of fentanyl was begun at a rate of 150 µg/min via the antecubital iv catheter.

Approximately 3.5 min after the infusion was begun, the patient began to have repeated jerking movements of his right hand and arm. Within seconds, similar movements were observed in the left arm and then in both lower extremities. The movements were rhythmic, with a frequency of approximately 100/min. These movements did not "march," and no tonic phase was observed. Muscles of the head and neck were not involved, and it was unclear if the trunk muscles were involved. The eyelids were passively opened, and no deviation of the eyes was seen. A succinylcholine drip was begun as soon as the abnormal movement started. Several minutes were required before the succinylcholine produced complete relaxation, yet, clinically, ventilation was assisted without difficulty.

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