Anesthetic and Dental Management of a Child With IMAGE Syndrome

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IMAGE syndrome (OMIM 300290) is a rare multisystem disorder that has a broad phenotypic presentation. Though variable, this disorder mainly consists of Intrauterine growth retardation, Metaphyseal dysplasia, Adrenal hypoplasia congenita, and Genital abnormalities. Patients with IMAGE syndrome present as an uncommon yet important challenge for dentists and anesthesiologists due to their wide range of dysmorphic facial features, adrenal insufficiency, electrolyte imbalances, and need for steroid replacement. The purpose of this case report is to describe the successful anesthetic management of a pediatric patient diagnosed with IMAGE syndrome who presented for full mouth dental rehabilitation.

Key Words: IMAGE syndrome; Adrenal hypoplasia; General anesthesia; Dental rehabilitation.

IIMAGE syndrome (OMIM 300290) is a rare multisystem disorder, first reported by Vilain et al¹ in 1999. The syndrome has a broad phenotypic presentation but is characterized by Intrauterine growth retardation, Metaphyseal dysplasia, Adrenal hypoplasia congenita, and Genital abnormalities.¹ Clinical manifestations of IMAGE syndrome are highly variable; however, all affected patients have some degree of primary adrenal insufficiency and require steroid supplementation.² In this case report, we describe the dental and anesthetic management of a young child with a diagnosis of IMAGE syndrome.

CASE REPORT

A 33-month-old girl with IMAGE syndrome, nephrocalcinosis, and a history of failure to thrive was referred to

our clinic by a private practice pediatric dentist for evaluation and treatment. The chief dental complaint was pain in the posterior of the patient's mouth from multiple severely decayed teeth. The patient initially presented to an outside hospital with an adrenal crisis, noted with low glucose, low sodium, and elevated potassium. Subsequently, she had been evaluated by endocrinology, genetics, orthopedics, nephrology, and ophthalmology. She exhibited the classical reported components of IMAGE syndrome including a history of intrauterine growth retardation, left hip dysplasia, and bilateral absence of adrenal glands.

According to the history obtained from the parents, their daughter was born prematurely at 36 weeks, hospitalized for 45 days, and on a ventilator for 6 weeks. She was small for her age at less than the fifth percentile for height and weight. Her head and facial features were dysmorphic with microcephaly and low set ears (Figure 1). In addition, she had global developmental delays. Her medications included hydrocortisone and fludrocortisone for her adrenal insufficiency. The patient was previously on sodium replacement therapy, but had

Received January 9, 2014; accepted for publication August 23, 2014.

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Anesth Prog 61:165–168 2014

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ISSN 0003-3006/14

SSDI 0003-3006(14)

Figure 1. Dysmorphic facial features of patient with IMAGE syndrome.

been weaned off prior to presentation. Her parents reported an egg allergy.

The clinical examination revealed full primary dentition with several abscessed teeth, gingival inflammation with poor oral hygiene, and crowding (Figure 2). The mandibular incisors were lingually inclined and conical, as was the primary maxillary right lateral incisor. Her palate was high, narrow, and constricted. There was a flush terminal plane relationship on both sides with 10% overbite and an overjet of 6 mm. Due to her young age, noncooperative behavior, complex medical history, and the extent of the dental care required, it was decided to complete her care in the operating room under general anesthesia.

On preoperative anesthesia evaluation, the patient's height (78.5 cm) and weight (7.7 kg) were both below the first percentile for age. The patient had a narrow facies, but her airway exam was within normal limits including size and shape of the chin, thyromental distance, and range of motion of her neck; her airway was Mallampati Class I. A preoperative electrolyte panel was checked and within normal limits.

The child was premedicated with 3.75 mg of midazolam oral syrup. Standard American Society of Anesthesiologists monitors were placed, and mask induction was uneventfully completed with oxygen, nitrous oxide, and sevoflurane (8%). A 24-g intravenous

Figure 2. High, narrow, and constricted palate with multiple carious nonrestorable teeth and evidence of dental abscesses.

catheter was placed in the right wrist after induction. The patient was paralyzed with vecuronium 0.7 mg to facilitate intubation. Although the patient had an initial direct laryngoscopy that revealed a grade I view, it was challenging to navigate both the blade and Magill forceps due to space constraints associated with the patient's high arched, narrow palate. The patient ultimately underwent an atraumatic nasal fiberoptic intubation with a 4.5-mm lubricated cuffed nasal RAE tube in the right nares. The throat pack was placed by the pediatric dentist.

The patient was given 250 mg of intravenous ampicillin, and the mouth was swabbed with 0.12% chlorhexidine gluconate prior to the dental procedure. The patient received a 40 mg intravenous stress dose of hydrocortisone at the start of surgery and tapering doses every 4 hours for the first day postoperatively, as recommended by the endocrinology team. Thereafter, her usual 2-mg dose of hydrocortisone was administered orally every 8 hours. For the duration of the procedure, the patient was maintained with an end-tidal concentration of 2.6% sevoflurane, supplemented by 1 mg of morphine after 2.5 hours, 0.25 mg of morphine near the end, and another 0.25 mg of morphine upon her arrival in the postanesthesia care unit. She remained normotensive with other vital signs also within normal limits of her baseline. Use of narcotics was minimal due to the nature of the dental treatment.

A full series of radiographs was taken at this time (Figure 3). After a thorough prophylaxis, a treatment plan was generated, and all carious primary teeth were either extracted or restored with reinforced glass ionomer resins, composite resins, or stainless steel crowns. Pulp therapy was necessary for several teeth. Ondansetron 0.5 mg was administered intravenously for prophylaxis against postoperative nausea and vomiting. Because the total duration of the surgical procedure was 182 minutes and no additional vecuronium was given

Figure 3. Full radiographic series.

after the initial dose, reversal of the neuromuscular blockade was not necessary. After the usual extubation criteria were met, the patient was extubated and transported to the postanesthesia care unit breathing spontaneously on room air. Her recovery was uneventful, and the patient was discharged in 1 hour to her hospital room. Routine follow-up via telephone at 24 hours and at the pediatric dentist's office at 4 weeks revealed no surgical or anesthetic complications.

DISCUSSION

Clinical manifestations of IMAGE syndrome are highly variable; however, since all patients with IMAGE syndrome have some degree of primary adrenal insufficiency, they are considered to be at risk for an acute adrenal crisis.³ Other common features in patients with IMAGE syndrome include craniofacial dysmor-

phisms and a myriad of skeletal abnormalities involving epiphyses and metaphyses most likely leading to their short stature.¹ Rarely are patients reported with bone dysplasia.³ Genital abnormalities are found only in male patients and often include micropenis, hypospadias, and cryptorchidism.^{1,3–6} Hypercalciuria,^{1,4} hypercalcemia,^{1,4} cleft palate,⁷ nephrocalcinosis,¹ failure to thrive,^{3,5–7} and sensorineural hearing loss⁷ have also been reported in association with this syndrome. The cause of IMAGE syndrome remains unknown, although various genes have been hypothesized to play a role in the syndrome. Some studies suggest an autosomal recessive mode of inheritance,^{4,5} whereas other studies propose inheritance through genomic imprinting of an autosomal gene.³ Our patient, with a history of intrauterine growth retardation, left hip dysplasia, and bilateral absence of adrenal glands, met the criteria for a diagnosis of IMAGE syndrome. In addition, she had dysmorphic facial features, microcephaly, nephrocalcinosis, global developmental delays, and remained below the fifth percentile for height and weight.

There are numerous anesthetic considerations for the patient with IMAGE syndrome. All patients have some degree of adrenal insufficiency due to adrenal hypoplasia congenita, which is a rare form of primary adrenal insufficiency that usually manifests during infancy or early childhood as hypoplastic or abnormal adrenal gland development. Untreated adrenal failure is incompatible with life. Regardless of the mode of inheritance, the approach to treatment is similar to that of most other disorders of adrenal insufficiency. Replacement glucocorticoid therapy is recommended for patients with adrenal hypoplasia congenita and IMAGE syndrome. The anesthesiologist needs to be prepared to manage and stabilize any electrolyte imbalances and blood pressure instability as a result of the adrenal hypoplasia congenita. These patients should be stabilized on medical therapy prior to presenting to the operating room. The therapy includes mineralocorticoid therapy, glucocorticoid therapy, or both, depending on the patient's presentation. An endocrinology consultation should be obtained prior to the patient's surgery. Manifestations of mineralocorticoid deficiency result in hyponatremia, hyperkalemia, and hypotension. These signs can be a consequence of failure of the adrenal glands to release adequate mineralocorticoids, but also can occur as a result of failure to release adequate glucocorticoids. This is because glucocorticoids possess mineralocorticoid properties as well, particularly inotropic activity. As a result, glucocorticoid replacement should be administered during periods of increased stress, such as severe illness, trauma, and surgery.⁸

There are other special considerations that should be taken into account in the management of patients who

are on long-term exogenous steroid therapy. Glucocorticoids are anti-inflammatory agents and have significant immunosuppressive effects. At physiologic levels, the effects of glucocorticoids help to regulate inflammation and maintain homeostasis. However, at high-dose long-term levels, the effects of exogenous glucocorticoids can cause significant immunosuppression, placing patients at a greater risk of infection.⁹ Thus, management of patients with adrenal insufficiency should include antibiotic prophylaxis before major dental procedures. Glucocorticoids are also important regulators of mineral metabolism and significantly influence bone formation and resorption.¹⁰ In glucocorticoid excess, decreases in bone mineral density occur as a result. The developing skeletons of children are especially vulnerable with cortical bone more sensitive than trabecular bone.¹¹ Thus, management of patients with adrenal insufficiency should include awareness of possible lower bone density in the head and neck and extra care during major dental procedures.

The management of patients with IMAGE syndrome must include attention to the wide range of craniofacial dysmorphisms that may complicate dental treatment and the potential difficulty of securing an airway. Classical facial features of IMAGE syndrome are frontal bossing, a broad nasal bridge, and low set ears.^{1,3,4,6} Few patients have also presented with a high-arched palate and craniosynostosis.^{1,4} Case reports have also indicated that microcephaly, micrognathia, a high arched palate or cleft palate, and thin lips are also common among patients.^{1,4–6,12} Radiographically, patients have often been found to have delayed bone age and maturation.^{1,3,4,12} By definition, patients with IMAGE syndrome have intrauterine growth restriction, and several continue to suffer with postnatal growth failure. This combination of features may lead to challenges in endotracheal tube sizing and placement. Scoliosis, although uncommon with the syndrome, can be severe when it is present and may further complicate securing the airway.^{5,6}

In maintaining the oral health of patients with IMAGE syndrome, a pediatric dentist must be aware of these craniofacial features as they may potentially present as a challenge during dental procedures. Additionally, a pediatric dentist should work in conjunction with other specialists such as orthodontists and oral surgeons in providing comprehensive care. Follow-up care is critical in these patients to reinforce prevention, monitor oral hygiene, guide growth and development, and maintain overall dental health. The patient with a rare syndrome such as IMAGE presents unique challenges for the anesthesiologist and pediatric dentist.

Since so little is known about IMAGE syndrome, we had no guidelines regarding the anesthetic or dental

management of this patient. Based on the literature, we knew that some of our main concerns would include electrolyte imbalances, steroid replacement, dysmorphic craniofacial features, and a potentially difficult airway. By having open communication with the family, anesthesiologist, endocrinologist, and pediatric dentist, we were able to successfully treat a young patient with IMAGE syndrome under general anesthesia for full mouth dental rehabilitation.

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