

ABSTRACT

The population of special needs patients in dental offices is growing. Therefore, the demand for well-trained, educated practitioners must increase to fit the need. Conditions such as intellectual developmental disorder, Down syndrome, and autism spectrum disorder are more readily encountered in dental settings. However, it is equally appropriate to identify management techniques for patients with less common conditions. A case is reported in which a 38-year-old Caucasian male with a history significant for acrometageria and associated signs of Mallampati Class IV, micrognathia, decreased mouth opening, decreased thyromental distance, and decreased cervical range of motion presented for routine dental treatment under intravenous sedation. Providers should recognize appropriate management techniques to safely and effectively care for a wide patient demographic.

KEY WORDS: special needs management, acrometageria, behavior guidance, sedation, dental treatment, general anesthesia

Management of patient with acrometageria for routine dental treatment: A case report

Andrew S. Young, DDS;^{1*} Matthew R. Cooke, DDS, MD, MPH;²
Lynne M. Taiclet, DMD³

¹Resident in Dental Anesthesiology, School of Dental Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania; ²Assistant Professor, Department of Dental Anesthesiology and Pediatric Dentistry, School of Dental Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania; ³Director and Assistant Professor, Center for Patients with Special Needs, School of Dental Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania.

*Corresponding author e-mail: any29@pitt.edu

Spec Care Dentist 36(1): 32-38, 2016

Introduction

Acrometageria is a progeroid (premature aging) syndrome characterized by a short, thin stature, beaked nose, atrophy on extremities and little subcutaneous fat.¹⁻³ Mode of inheritance is yet to be determined, although both acrogeria and metageria have been identified as autosomal recessive.

Acrometageria was first described by Greally *et al.* in 1992 as a manifestation of two premature aging syndromes—acrogeria and metageria.³ While acrogeria was described initially by Gottron (1940) who reported on siblings with skin changes characteristic of aging on their hands and feet, metageria was described by Gilkes *et al.* (1974) as a phenotype differing sufficiently to warrant a separate classification.^{4,5} In 1985, Kaufman *et al.* explored the occurrence of acrogeria and metageria in the same family leading authors to suggest that the two conditions exist as variants of one another.⁶ These observations have led to further suggestions proposing the existence of a spectrum of disorders including both acrogeria and metageria referred to by an encompassing term—acrometageria.³ Due to their rarity, the aforementioned premature aging syndromes are largely defined by their clinical manifestations. A comparison of acrogeria, metageria,

and acrometageria are outlined in Table 1.

In dentistry for special needs patients, practitioners encounter various conditions. Understanding how to manage patients with unique conditions is important for providing successful comprehensive dental care. By recognizing management options, both behavioral and pharmacological, dental providers may address barriers to treatment.

Case report

Medical history

A 38-year-old Caucasian male (162 cm, 51 kg) presented at the University of Pittsburgh School of Dental Medicine for comprehensive dental treatment in the Center for Patients with Special Needs. The patient had a medical history significant for acrometageria, scoliosis, cataracts, Raynaud's syndrome, and intellectual

Table 1. Clinical manifestations of premature aging syndromes.

	Acrogeria	Metageria	Acrometageria/This Patient
Stature	Normal	Tall and thin	Short and thin
Face	Micrognathia & atrophy of skin on tip of nose	Beaked nose	Micrognathia & beaked nose
Skin	Atrophy on extremities	Atrophy on limbs	Atrophy on extremities
Scalp hair	Normal	Fine and thin	Fine and thin
Eyes	Normal	“Prominent”	Normal
Nails	Dystrophic and thickened	Normal	Normal
Limbs	No leg ulcers	Generalized loss of subcutaneous fat	Little subcutaneous fat
Cardiovascular	Normal	Early atherosclerosis	Normal
Skeletal	Normal	Normal	Scoliosis
Metabolic	Normal	Early onset diabetes mellitus	Normal
Inheritance	Autosomal recessive	Autosomal recessive	Unknown
Sex incidence	Mostly female	Both equally	Male
Onset	Birth	Birth	Birth
Prognosis	Normal lifespan	Dependent on severity of DM and atherosclerosis	Unknown

Table 2. Patient information.

Patient demographic	38-year-old Caucasian male
	162 cm, 51 kg
	ASA II
Medical history	Acrometageria
	Scoliosis
	Cataracts
	Raynaud’s syndrome
	Intellectual disability
Allergies	Penicillin
Medications	Denies
Past surgical history	Orthopedic Hand Surgery for Joint Hypermobility 1993
	Spondylodesis C3/C4 following Motor Vehicle Accident 2004
Social history	Denies
Family history:	Mother: Factor V Leiden Thrombophilia
	Father: None

disability with associated signs of hyperflexible joints, thin stature, beaked nose, atrophy on extremities, and little subcutaneous fat (Table 2). Further physical examination and airway assessment revealed Mallampati Class IV, micrognathia, decreased maximum mouth opening, decreased thyromental distance, and decreased cervical range of motion;

these predictors of difficult airway potentially complicate sedation and/or general anesthetic management (Figure 1).

The patient’s father mentioned previous surgical history for hyperflexible joints of the patient’s hands in 1993. A traumatic motor vehicle accident (MVA) resulted in a compound fracture of his right humerus and fracture of cervical

vertebrae (C3/C4) which was corrected by spondylodesis in 2004 (Figure 2). The remainder of the patient’s information including patient demographic, past medical history, allergies, medications, social history, and family history are outlined in Table 2.

Dental history

The patient exhibited shy and moderately anxious demeanor, but was largely cooperative for the dental examination with verbal and physical (handholding) encouragement from the patient’s father. Frequent breaks were required due to the patient’s limited opening necessitating shorter appointments for subsequent visits.

The initial oral examination revealed existing restorations on #30 and #31 with recurrent caries, which were treatment planned for amalgam restorations. Subsequent dental appointments revealed secondary caries on #14MO and #31MO; primary caries on #30MO, #15O, #21D, #28D, and #31B; and incipient caries on #5D, #19M, and #21O. Tooth #14 was treatment planned for amalgam, #31 for MOB amalgam, and remaining caries for resin-based composite restorations (Table 3). At a subsequent restorative appointment, tooth #14 restoration was

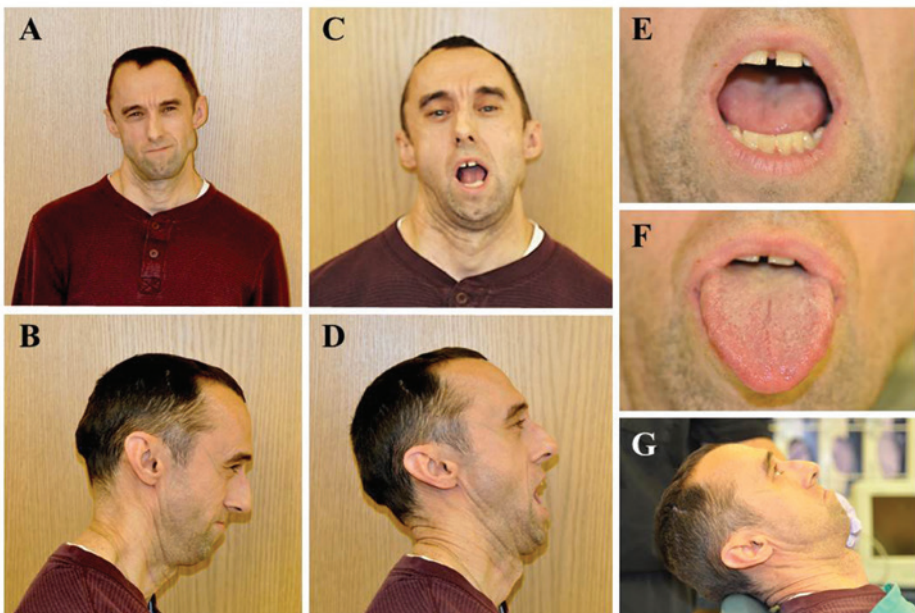


Figure 1. Physical examination and airway assessment as predictors of difficult airway. (A) Frontal view. (B) Sagittal view. (C) and (E) Frontal view of patient exercising maximum opening. The decreased mouth opening may be appreciated. (D) Sagittal view of patient depicting decreased maximum opening and cervical range of motion. (F) Patient Mallampati Class IV. Decreased opening and limited visualization of soft palate, uvula, and tonsillar pillars. (G) Patient thyromental distance approximately two fingerbreadths.



Figure 2. (A) and (B) demonstrate the wrinkled skin on the hands and feet; hypermobile joints. (C) Incision site for spondylosis of cervical vertebrae C3/C4 following traumatic MVA.

determined to be unrestorable and treatment planned for extraction. The patient was successfully managed for initial

examination, radiographs and subsequent restorative appointments with behavioral, psychological, and other

nonpharmacological techniques. Tooth #14 was planned for extraction under intravenous sedation.

The patient's father accompanied him to the treatment area and provided encouragement for intravenous peripheral catheter placement. A 20-gauge catheter was secured in the left forearm, through which the patient received midazolam 2 mg and sufentanil 2.5 mcg. Propofol infusion was initiated at a rate of 40 mcg/kg/min and supplemented with incremental dexmedetomidine in 4 mcg doses over the course of treatment for a total of 24 mcg. The patient's father remained in the treatment area until the patient was sedated. The patient received 2% lidocaine (72 mg) with epinephrine 1:100:000 (0.036 mg) for maxillary infiltration and tooth #14 was extracted with minimal bleeding. Following extraction, the propofol infusion was discontinued; sedation was maintained with dexmedetomidine and a second dose of sufentanil 2.5 mcg. The resin restoration of tooth #21 was tolerated with minimal-to-moderate sedation. Based on limitations associated with providing dental care in a learning institution and the patient's disabilities, the remaining treatment was scheduled to be completed during subsequent appointments.

Discussion

Progeroid syndromes are conditions characterized by premature aging and shortened life expectancies.² Frequently, patients exhibit signs of accelerated aging such as sclerotic skin, heart and bone abnormalities, and growth attenuation. Affected individuals display respiratory and cardiovascular capacities comparable to the elderly.⁷ In many cases, craniofacial features are evident including "bird-like" facies, alopecia, craniofacial disproportion, hypodontia, delayed eruption, and dental crowding. While there are over a dozen syndromes that tend to be associated with premature aging, the approach to dental management of patients is relatively similar.²

In patients with progeroid syndromes, attention to developmental status as it relates to chronological age is

Table 3. Patient's dental treatment plan.

Tooth #	Existing restorations	Existing defects	Treatment plan
5	None	D incipient caries	No treatment
14	MO amalgam	MO secondary caries	MO amalgam
15	None	O primary caries	O resin-based composite
19	None	M incipient caries	MO amalgam
21	None	O incipient caries	O resin-based composite
28	None	D primary caries	D resin-based composite
30	DO amalgam	M Caries, O secondary caries	MOD resin-based composite
31	MO amalgam	B primary caries. MO secondary caries	MOB amalgam

Note: Tooth #14 was eventually retreatment planned for extraction.

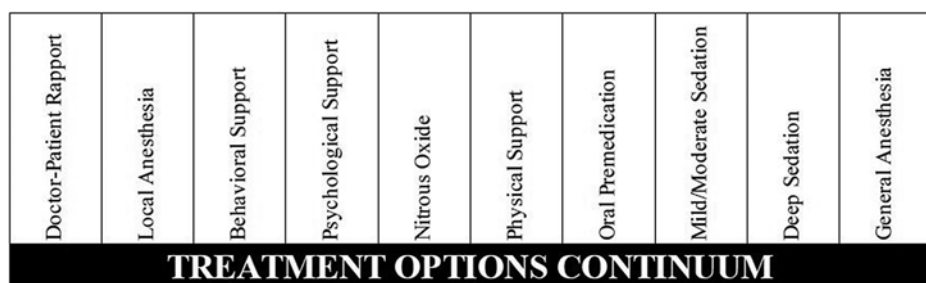


Figure 3. Patient management options.

Table 4. Patient's conditions and associated limitations.

Patient's condition	Patient's associated signs and symptoms	Considerations for dental treatment
Dental anxiety	Shy, restrained demeanor	Duration of procedure
	Moderate fear of dental treatment	Doctor-patient communication
Acrometageria	Hyperflexible joints	Patient positioning
	Mallampati Class IV	Duration of procedure
	Micrognathia	Inability to sustain mouth opening
	Decreased mouth opening	
	Decreased cervical range of motion	
Scoliosis	Spinal deformity/curvature	Patient positioning
		Duration of procedure
Intellectual disability	Limitations in adaptive functioning	Chronological versus developmental age
		Doctor-patient communication
Trauma/surgical history	C3/C4 spinal fracture and spondylolysis	Patient positioning
		Duration of procedure

important for successful treatment. A thorough medical history and clinical examination are useful in establishing

parameters for care. Scheduled appointment times, length of appointments, stress reduction protocols, and proper

positioning are other necessary considerations.⁸ As with older adults and geriatric patients, those with premature aging may have limitations which create a need for specialized dental care.

The American Dental Association (ADA) defines patients with special needs as “those who due to physical, medical, developmental, or cognitive conditions require special consideration when receiving dental treatment.”⁶ A variety of management techniques have been developed to aid patients with special needs to receive dental treatment services (Figure 3). Clinical guidelines have been drafted by the American Academy of Pediatric Dentistry (AAPD) and supported by the Special Care Dentistry Association (SCDA).⁹ Furthermore, in 2009, the SCDA published a consensus statement on sedation, anesthesia, and alternative techniques for people with special needs.¹⁸ These techniques and considerations were applied in treatment of the patient presented in this case report.

Information gathered from the comprehensive oral evaluation was utilized for proper patient assessment—considering medical history, physical examination, and dental treatment plan.⁹ In determining a treatment modality for this patient, consideration was given to each of the patient's conditions and associated limitations to treatment (Table 4).

For the initial comprehensive oral examination, diagnostic radiographs, adult prophylaxis, and restorative treatment, the patient was successfully

Table 5. Behavioral support techniques.

Behavioral support techniques	Description
Voice control	Alteration of vocal volume, pace, and intonation to gain patient's attention and influence behavioral direction
Nonverbal communication	Recognition of nonverbal cues: sensitivity to touch, body language, and facial expressions
Tell-show-do	Approach of adding sensory demonstration cues (visual, auditory, touch, proprioception, etc.) to a simple verbal description of procedure prior to the performance of procedure
Positive reinforcement	Process of rewarding acceptable/desired behavior with verbal praise, expression, touch, or tokens
Contingent escape	Offer of momentary cessation of treatment or other positive reinforcers, conditional upon periods of acceptable target behavior
Distraction	Method of diverting a patient's mental focus to positive thoughts, favorable environmental stimuli, or other stimulating sensory images in an effort to override unpleasant procedures
Parental presence	Utilization of a parent to increase the patient's psychological comfort and reduce patient anxiety

Source: Lyons (2009)

Table 6. Keys to patient assessment.

Evaluation factors
1. Patient's health history and current medical status
2. Likelihood of procedures being completed successfully
3. Time and effort required from the patient and health care provider
4. Cost of contemplated procedures
5. Risk of side effects of any aspect of treatment
6. Social environment and support available
7. Availability of various treatment modalities
8. Urgency of care for that individual

Source: Glassman *et al.* (2009)

managed with behavioral, psychological, and other nonpharmacological support techniques. Behavioral support techniques utilized in patient treatment included voice control, nonverbal communication, tell-show-do, positive reinforcement, contingent escape, distraction, and parent presence.¹² The patient exhibited moderate anxiety, but appeared to benefit from the presence of his father at chairside for the duration of the procedure. The father assisted by providing periodic verbal and physical encouragement in the form of reassurance and handholding, respectively. The patient's limited opening and micrognathia necessitated frequent rest periods during procedures. Scoliosis, limited

range of motion, previous history of trauma, and intellectual disability affected the patient's ability to tolerate long appointments requiring shorter sessions. Table 5 details the behavioral support techniques used with this patient.

Psychological support was also utilized in the treatment of this patient. Current literature shows that patients with special needs are sometimes more comorbidly dental phobic than others.¹³ The patient presented in this case report appeared to experience moderate dental anxiety, which was addressed with psychological support techniques including relaxation, breathing, and effective use of developmentally appropriate operatory

language. These methods created a calm work setting for both doctor and patient. Social support was additionally employed to provide continuity of treatment through patient and family education. This approach benefited the patient with knowledge for home care and overall oral health maintenance.¹¹

Nonpharmacological techniques proved successful for examination, radiographs, and operative dentistry; however, deep sedation was provided for a potentially difficult extraction of tooth #14. Patients with special needs require a variety of modalities for treatment, ranging from good rapport to general anesthesia.

While the ADA and AAPD provide a number of guidelines in literature related to providing sedation and general anesthesia for dentistry, few specifically discuss indications for use of sedation or anesthesia. The focus of guidelines is often on education for methods of delivery.^{9,10,14-16} The Special Care Dentistry Association lists seven rationales for use of sedation and general anesthesia, which were similar rationales for treatment of the presented patient.^{17,18} The factors include: healthy history, medical status, time and effort, cost, and risks among others listed in Table 6.

The patient presented fulfilled a number of rationales for sedation and general anesthesia. He showed decreased developmental status versus chronological age, required a reduction in pain and anxiety, and was not able to cooperate in a manner that would allow surgery to be completed in an optimally safe fashion. The patient also requested the use of sedation or general anesthesia for extraction of #14, recognizing the utility of decreasing pain and anxiety.

In the decision-making process for determining treatment planning sedation/general anesthesia, a number of factors are considered (Table 7).¹⁶ The presented patient's health history and physical status played a major role in forming a definitive plan for combination deep and moderate sedation. Manifestations of patient's acrometageria revealed Mallampati Class IV, micrognathia, decreased mouth opening, decreased thyromental distance, and decreased cervical range of motion; all are

Table 7. Rationales for use of sedation and general anesthesia.

1. Individuals with cognitive impairment or emotional conditions who have difficulty understanding what is expected in a dental treatment situation
2. Patient whose fear about receiving dental treatment prevents them from receiving the needed treatment
3. Patients who are unable to sit in a dental chair or remain still enough to have dental procedures performed
4. Patients who have extensive dental needs that would require extended dental treatment over a prolonged period of time
5. Patients who require dental procedures that cannot easily be performed with local anesthesia because of an inability to achieve adequate local anesthesia for that procedure
6. Individuals with complex medical problems who require intra- and perioperative monitoring
7. Individuals with complex medical problems (e.g., severe hypertension and cardiac or respiratory disease) whose physiologic state will be more safely controlled in a sedated or anesthetized state
Source: Glassman <i>et al.</i> (2009)

Table 8. Advantages and disadvantages of general anesthesia.

Advantages	Disadvantages
1. Success rate is 100%	1. The patient is unconscious during treatment
2. Patient cooperation is not absolutely necessary	2. The patient's protective reflexes are depressed
3. The patient is unconscious during treatment	3. Vital signs are depressed
4. The patient does not respond to pain	4. Advanced training is required for the person administering GA
5. Amnesia is present after procedure	5. A team of professionals (rather than an individual dentist) is required to provide treatment
6. General anesthesia may be the only technique that will prove successful for certain patients	6. Special anesthesia equipment is required
7. The onset of action of general anesthesia is usually quite rapid	7. A recovery area must be available for postoperative monitoring (a private setting or the dental operatory itself may serve as a recovery room)
8. Titration is possible with the patient receiving smallest volume of drug required	8. Intraoperative and postoperative complications are more common during GA than sedation
	9. It is recommended that patients receiving GA should refrain from drinking clear liquids for at least 2 hours preoperatively and solids and nonhuman milk for at least 6 hours preoperatively
	10. Patients receiving GA may require extensive preoperative evaluation and testing
Source: Malamed (2010)	

indications of a poor airway. Furthermore, intubated general anesthesia may have proven difficult given the patient's history of trauma, fusion of C3 & C4, scoliosis, hyperflexible joints, predisposition to fractures, and concerns with patient head and

neck positioning for endotracheal tube placement.

After considering the risks and benefits of providing sedation and general anesthesia (Table 8), combination therapy of deep sedation with transition

to minimal/moderate sedation was selected. The patient was properly positioned with a shoulder roll and mild chin support to maintain airway patency for extraction. The dental history demonstrated cooperation at previous visits without sedation for routine restorative dentistry; a reduction in sedation would likely be tolerable for the remainder of the patient's appointment. Behavioral and psychological supports were used in conjunction with minimal-to-moderate sedation for completion of the appointment without incident. In fact, literature shows that good communication has the potential to facilitate the acceptance of successful pharmacosedation.¹⁹

The remaining restorative treatment was planned for completion with non-pharmacological support techniques. In the event nonpharmacological techniques prove unsuccessful, sedation may be reconsidered. Combination support proved to be the most effective approach for subsequent appointments. However, in addition to understanding support techniques, recognizing barriers to care is also important for comprehensive treatment of dental patients with special needs.²⁰ Access to proper health care is hindered by insufficient numbers of trained personnel, difficulty in obtaining informed consent for treatment decisions, lack of accessibility to facilities, and inadequate financial coverage. Successful oral health care support for patients with disabilities requires identification of patient management techniques within the larger scheme of addressing general barriers to treatment.

Conclusion

The population of special needs patients is growing with advances in medical care, deinstitutionalization, and changing societal values.¹¹ Many of these individuals require additional attention and support for dental treatment. While a number of treatment modalities exist, universal guidelines for their implementation are not always clear. However, with proper patient assessment, provider education, and treatment planning consideration, a combination of appropriate support

modalities may be utilized in successfully caring for patients with special needs.

References

1. Brown WR. Progeroid syndromes. In: *The Encyclopedia or Aging: A Comprehensive Resource in Gerontology and Geriatrics*. 2nd ed. New York, NY: Springer Publishing Company; 1995: 765-7.
2. Fossel M. The progerias. *J Anti-aging Med* 2003;6(2):123-38.
3. Greally JM, Boone LY, Lenkey SG, Wenger SL, Steele MW. Acrometageria: a spectrum of premature aging syndromes. *Am J Med Genet* 1992;44:334-9.
4. De Groot WP, Tafelkruyer J, Woerdeman MJ. Familial acrogeria (Gottron). *Br J Dermatol* 1980;103:213-23.
5. Gilkes JJH, Sharvill DE, Wells RS. The premature ageing syndromes report of eight cases and description of a new entity named metageria. *Br J Dermatol* 1974;91:243-62.
6. Kaufman I, Thiele B, Marle G. Simultaneous occurrence of metageria and Gottron's acrogeria in one family. *Z Hautkr* 1985;60: 975-84.
7. Domingo DL, Trujillo MI, Council SE, et al. Hutchinson-Gilford progeria syndrome: oral and craniofacial phenotypes. *Oral Dis* 2009;15(3):187-95.
8. Little JW, Falace DA, Miller CS, Rhodus NL. Dental management of older adults. In: *Dental Management of the Medically Compromised Patient*. 7th ed., Chapter 30. St. Louis, MO: Mosby; 2012: 545-47. Print.
9. American Dental Association. American Dental Association guidelines for the use of sedation and general anesthesia by dentists. Adopted by the American Dental Association House of Delegate; October 2007: 1-14.
10. American Academy of Pediatric Dentistry. Guideline on management of dental patients with special health care needs. Adopted by the American Academy of Pediatric Dentistry Council on Clinical Affairs; 2012: 161-6.
11. Glassman P, Miller C. Social supports and prevention strategies as adjuncts and alternatives to sedation and anesthesia for people with special needs. *Spec Care Dentist* 2009;29(1):31-8.
12. Lyons R. Understanding basic behavioral support techniques as an alternative to sedation and anesthesia. *Spec Care Dentist* 2009;29(1):39-50.
13. Peltier B. Psychological treatment of fearful and phobic special needs patients. *Spec Care Dentist* 2009;29(1):51-7.
14. American Dental Association. Guidelines for teaching pain control and sedation to dentists and dental students. Adopted by the American Dental Association House of Delegates; 2012: 1-17.
15. American Academy of Pediatric Dentistry. Guideline on the elective use of minimal, moderate, and deep sedation and general anesthesia for pediatric dental patients. *Ped Dent* 2005;27(7 Reference Manual):110-8.
16. Dougherty N. The dental patient with special needs: a review of indications for treatment under general anesthesia. *Spec Care Dentist* 2009;29(1):17-20.
17. Glassman P. A review of guidelines for sedation, anesthesia, and alternative interventions for people with special needs. *Spec Care Dentist* 2009;29(1):9-16.
18. Glassman P, Caputo A, Dougherty N, et al. Special care dentistry association consensus statement on sedation, anesthesia, and alternative techniques for people with special needs. *Spec Care Dentist* 2009;29(1):2-8.
19. Malamed SF. *Sedation: A Guide to Patient Management*. 5th ed. St. Louis, MO: Mosby Elsevier; 2010: 428-31.
20. Raposa KA, Perlman SP. *Treating the Dental Patient with a Developmental Disorder*. Danvers, MA: Wiley-Blackwell; 2012: 83-95, 169-93.