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Rett syndrome—a rare and often misdiagnosed syndrome: case report

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Abstract

Rett syndrome is a recently discovered, progressive neurological disorder that occurs exclusively in females early in life. Females with this rare and often misdiagnosed clinical entity have unusual but distinctive oral/digital habits. Dentists aware of Rett syndrome and its distinct manifestations will be able to aid in early diagnosis and treatment of those afflicted.

A case report of a 4-year, 3-month-old white female with Rett syndrome and review of pertinent literature are presented. The previously reported oral/digital habits and oral manifestations of Rett syndrome are listed along with other unreported oral findings. Many of these oral manifestations are not unique to Rett syndrome. There is a likelihood that they may be related to trauma secondary to the excessive oral/digital habits, poor gait, and seizure activity.

Rett syndrome is a progressive neurological disorder that occurs exclusively in females and is characterized by normal early development followed by a progressive loss of acquired functions.

The purpose of this paper was to review this rare syndrome and increase awareness of the syndrome's manifestations among dental professionals, thereby aiding early diagnosis and treatment.

A case report of a 4-year, 3-month-old white female with Rett syndrome and review of pertinent literature are presented. The previously reported oral/digital habits and oral manifestations of Rett syndrome are listed along with other unreported oral findings.

The International Rett's Syndrome Association has noted that Rett syndrome often is misdiagnosed because there appears to be a lack of awareness of the syndrome by the professional.

Literature Review

Diagnostic Criteria and Characteristic Features

Rett syndrome first was described by Rett (1966). He reported a progressive neurological disorder that occurred exclusively in girls and which had its onset near the end of the first year of life or during the first 6 months of the second year of life. Characteristic features included autistic behavior, dementia, apraxia of gait, loss of facial expression, and stereotyped use of the hands.

Leiber and Olbrich (1972) first described this condition in detail in their catalog of syndromes as "Rett syndrome." Hagberg et al. (1983) brought the syndrome to the attention of neurologists worldwide when they described 35 cases from Sweden, France, and Portugal with a uniform and striking progressive encephalopathy. Hagberg (1985) published the first diagnostic criteria for Rett syndrome (Table 1).

One year later, Al-Mateen et al. (1986) summarized the characteristics of Rett syndrome into 3 phases (Table 2, next page) and reinforced the previous findings of Rett (1966) and Hagberg et al. (1983). They also identified 2 previously unreported features of Rett syndrome: an extrapyramidal disorder and lactic acidemia.

The first recognized cases of Rett syndrome in the United States were reported by Holm in 1985. Murphy

TABLE 1. Rett Syndrome: Diagnostic Criteria for Inclusion

1. *Sex* – Female
2. *Normal prenatal and perinatal period* – Essentially normal psychomotor development through the first 6, often 12–18 months of life
3. *Normal head circumference at birth* – Deceleration of head growth (and therefore, by inference, brain growth) between ages 6 months–4 years.
4. *Early behavioral, social, and psychomotor regression* – Loss of achieved abilities, evolving communication dysfunction, dementia
5. *Loss of acquired purposeful hand skill* – Through ages 1–4 years
6. *Hand wringing-clapping-"washing hand" stereotypes* – Appearing between ages 1–4 years
7. *Appearance* – Gait apraxia and truncal apraxia/ataxia appearing between ages 1–4 years
8. *Diagnosis* – Tentative until 3–5 years of age.

(Hagberg et al. 1985)

TABLE 2. Characteristic Features of Rett Syndrome

First Phase

- Stagnation of development acquisitions (4–18 months of age)
- Insidious, occasionally acute, regression (18–36 months of age)
- Impairment or loss of speech
- Autistic behavior with distinctive hand movements (hand-washing automatism)
- Frequent hyperventilation

Second Phase

- Minor motor seizures (onset at 2–4 years of age)
- Loss of purposeful use of extremities (apraxia)
- Extrapyramidal symptoms (choreoathetosis and dystonia in 50% of cases)

Chronic Phase

- Progressive muscle wasting and spasticity
- Plateau of head circumference (as early as 1 year of age)
- Plateau of weight and usually height (after 10 years of age)

(Al-Mateen et al. 1986)

et al. (1986) stated that Rett syndrome: has been identified in 4 of the 7 continents; is seen across all social, religious, and ethnic groups; and has eluded all efforts to unravel its suggested genetic cause. Budden (1986) wrote that individuals with Rett syndrome frequently have been misdiagnosed as having mental retardation, cerebral palsy, and, most commonly, autism. Hagberg and Witt-Engerstrom (1986) stated that Rett syndrome can be misdiagnosed as infantile autism, childhood psychosis, spinocerebellar degeneration, neuraxonal dystrophy, hypotonic or ataxic cerebral palsy, minor motor epilepsy, toxic encephalitic process, infantile ceroid lipofuscinosis, phenylketonuria, tuberous sclerosis, mental retardation, heredoataxias, or leukodystrophies.

Important clinical criteria that aid in the syndrome's diagnosis include autism, loss of hand use and language. The most frequent misdiagnosis of Rett syndrome is infantile autism (Budden 1966). Kerr and Stephenson (1985) recommend that Rett syndrome should be considered whenever girls are referred with a diagnosis of autism. Naidu (1986) concludes that the loss of hand use and language before the age of 2 years are the most outstanding early clinical indices of suspicion.

Other clinical findings in children with Rett syndrome include: a shift from left-handedness to right-handedness near age 7 (Nomura et al. 1984; Olsson and Rett 1986); a possible defect in the specialized units of the inferior colliculus or higher auditory centers which subserve frequency sweep discrimination (Lenn et al. 1986); self-injurious hand biting (Iwata et al. 1986); stereotypic hand washing; apnea during active wakefulness, causing episodes of severe cerebral hypoxia (Cirignotta et al. 1986); and an abnormal electroencephalogram (EEG) portraying rhythmical slowing (Niedermeyer et al. 1986).

Olsson and Rett (1987) speculate that the characteristic hand washing stereotypies are due to a regression of acquired cognitive skills to the fourth stage of sensorimotor intelligence as described by Piaget (1952). These automatisms are a result of severe mental and motor regression and fulfill Piaget's description of primary circular reactions. Lenn (1986) felt that the stereotypic hand washing is most suggestive of involvement of the basal ganglia.

Etiology and Pathogenesis

The etiology of Rett syndrome is unknown (Bachmann et al. 1986; Harris et al. 1986; Nomura and Segawa 1986). Rett (1966) originally attributed the disorder to hyperammonemia, but this symptom is no longer essential in the diagnosis. The condition resembles a metabolic disorder; however, laboratory and metabolic studies have revealed inconsistent results.

During Rett syndrome's 23-year-old history, many other possible etiologies have been theorized. Holm (1985) speculated that the syndrome's etiology may be a new mutation caused by the environment. Hagberg et al. (1983) speculate that the disorder is caused by a dominant mutation on an X chromosome, which is lethal in males.

Another hypothesis of the pathogenesis of Rett syndrome suggests that symptoms result from an abnormality in the dopamine system, a neurotransmitter system which regulates the control of voluntary movements in the extrapyramidal system (Harris et al. 1986). Nomura et al. (1985) speculate that as Rett syndrome progresses, the dopamine system becomes hyperactive due to postsynaptic supersensitivity caused by hypoactive dopamine neurons.

Nomura and Segawa (1986) have clinical and polysomnographic data that support the hypothesis that Rett syndrome is due to a disorder of the noradrenergic, serononergic, and dopaminergic systems arising in the locus ceruleus, raphe nuclei, and substantia nigra, respectively.

The incidence of the condition is approximately 1 in 15,000 live female births (Kerr and Stephenson 1985; Hagberg 1985). Budden (1986) diagnosed 13 cases within a 9-month period and therefore suggests that the frequency of the disorder is higher. Moser (1986) stated there have been approximately 605 "classic" cases of Rett syndrome reported in the world, and of these, approximately 190 have been diagnosed in the United States. Life span appears to be unaffected although life tables have not yet been established; however, the

pathogenesis of this syndrome is progressive (Naidu et al. 1986).

Treatment

Treatment of the syndrome includes physical and occupational therapy to develop and maintain muscle tone, function, and contact (Hanks 1986). Music therapy has been used to lengthen attention span (Hanks 1986; Wesecky 1986). Various medications (ethosuximide, clonazepam, metharbital, ACTH, ketogenic diet, phenobarbital, phenytoin, clorazepate, valproate, and especially carbamazepine) have been prescribed for seizure control (Budden 1986; Haas et al. 1986; Philippart 1986).

Oral Manifestations

The oral manifestations of Rett syndrome have been reported only in the medical literature. The previously reported oral manifestations and oral/digital habits include: bruxism, drooling, hypersalivation, micrognathia, abnormal chewing, hand biting, spitting, tongue protrusion, narrow maxillary arch, high arched palate, stereotypic quick mouthing of objects using both hands, biting or rubbing of the dorsum of the hand against the teeth and lips, and digital-hand sucking or licking occurring between stereotypic hand washing episodes (Table 3).

Case Report

Medical History

A 4-year, 3-month-old white female was referred to the Riley Child Development Center by her pediatrician for a developmental evaluation due to concerns regarding hypotonia, developmental delays, and a gradual regression in some self-help skills and behavior.

The patient was born to a 26-year-old white female at full term following an uncomplicated pregnancy, 9-hour labor, and delivery. The mother reported no drug abuse, drug intake, smoking, or alcohol abuse during pregnancy. Apgar scores were 9 at 1 min and 10 at 5 min. The patient's birth weight was 9 pounds, 2 ounces. Other than a transient jaundice, there were no complications during the neonatal period and the patient was discharged from the hospital at 4 days of age with her mother. The mother became concerned with her daughter's health due to the child's decreased muscle tone at 6 months of age.

TABLE 3. **Rett Syndrome**

Oral Manifestations	*Oral Digital Habits*
• Abnormal chewing	• Handbiting
• Bruxism	• Drooling, spitting
• Hypersalivation	• Biting or rubbing of the dorsum of the hand against the teeth and lips
• Micrognathia	• Stereotypic quick mouthing of objects using both hands
• High arched palate	• Digital-hand sucking, licking, or wetting with saliva between stereotyic hand-washing episodes
• Narrow maxillary arch	
• Tongue protrusion	

(Adapted from Adkins 1986; Budden 1986; Hagberg 1986; Hanks 1986; Holms 1986; Iwata 1986; Moser 1986; Naidu 1986; Olsson and Rett 1987; Opitz 1986; Philippart 1986; Zappella 1986)

The child's developmental milestones were as follows: she rolled over at 5 months; sat with support at 5 months; scooted at 6 months; sat without assistance at 12 months; first began to use words at 12 months; crawled at 13 months; pulled to a stand at 16 months; walked by holding onto objects at 22 months; and walked independently at 29 months. Presently, she is not toilet trained and her speech has not progressed beyond the use of single words. By report, she attained an expressive vocabulary of approximately 75 words at 33 months of age. Since that time she has demonstrated a gradual but dramatic loss in her communication skills, and is currently using fewer than a dozen words meaningfully. At 33 months of age she was able to grasp with both hands and transfer between hands, but had not developed any clear hand preference.

The patient was evaluated at age 2 years, 10 months. The results of this evaluation, using the Gesell Scale of Development, indicated that the patient's adaptive functioning was at the 15-month level. She was able to follow simple commands, but demonstrated no verbalizations. The impression formulated at the time indicated that she was experiencing gross developmental delays and spastic encephalopathy with mild hypotonia.

She then was evaluated at the chronological age of 3 years. The results of this evaluation using the Stanford-Binet Intelligence Scale indicated that she was functioning with an IQ of 66.

The patient was evaluated at 4 years of age. The pediatrician's impression was that the patient was hypotonic with developmental delay. She had no known allergies and was not taking any medications. A CT scan and chromosome analysis were within normal limits. There was evidence of speech pattern regression and the development of behavior problems.

A 30-day follow-up visit with the pediatric neurology department revealed that she had severe behavior problems and possible autism. She displayed screaming fits that would last from several minutes to 3 hr, was violent, and would bite/hit those around her. Her mother reported that her daughter was talking less now than she did a year ago. Since 14 months of age,

there has been a dramatic and progressive loss of functioning in all domains.

The most recent evaluation was done at Riley Child Developmental Center in Indianapolis, Indiana, when the child's chronological age was 4 years, 3 months. The physical examination showed the patient's height and weight to be in the 75th percentile (National Center for Health Statistics). Positive clinical findings included hyperventilation, stereotypic hand washing activity, autistic-like features, mild to moderate generalized hypotonia with increased range of motion in the upper extremities, mild decreased range of motion in the hips (right more than left), mild tightness in the Achilles tendons and hamstrings bilaterally, symmetrical deep tendon reflexes, bilateral planovalgus deformity of feet, and clumsy gait without ataxia (Fig 1). Occipitofrontal head circumference was unchanged (50.5 cm) for the past 2 years. The patient's mother related symptoms resembling minor motor seizures. An EEG revealed right parietal temporal spikes. The audiometric exams revealed responses to sound field stimuli within normal limits; tympanograms were normal.

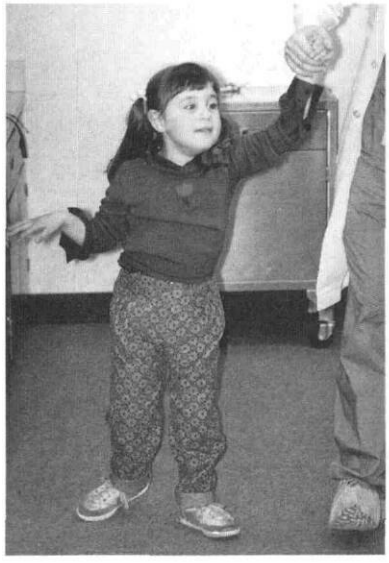

Fig 1. A pose of the stereotypic clumsy gait seen in Rett syndrome.

The results of this multidisciplinary developmental evaluation indicate that the patient is functioning in the severe to profound range of mental handicap. Previous test data and current findings indicate she has displayed a dramatic loss of functioning in all domains during the past 3 years. A number of unusual behaviors were noted: stereotypic hand washing, hyperventilation, full body rocking, and poor oral-motor control.

Dental History

The patient has been on a preventive dental program with a pediatric dentist since the age of 3 years. She has received an examination and prophylaxis on a routine basis. The mother was unaware of any dental problems. The patient lives in a community with a fluoridated water supply. The patient's mother brushes her child's teeth 3 times per day with an ADA-accepted fluoridated toothpaste.

Extraoral evaluation revealed a convex facial profile. The head, neck, face, and lips were within normal limits. The patient's dorsum of her right hand had a discolored, hyperker-atotic area (Fig 2).

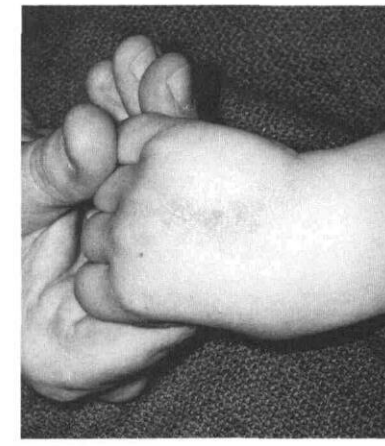

Fig 2. Hyperkeratotic area on dorsum of hand due to stereotypic handwashing.

Intraoral examination of the oropharynx, tongue, floor of the mouth, buccal mucosa, frena, gingiva, and periodontium were within normal limits. Palatal shelving was present (Fig 3).

Intraoral examination of the hard tissues revealed a bilateral distal step terminal plane, a Class II canine relationship, 4 mm of overjet, 100% overbite, a normal midline, primate spaces, no mandibular shift, and adequate maxillary and mandibular arch length. The eruption sequence of the primary dentition appeared to be within normal limits; permanent teeth were unerupted. There was no evidence of cross-bites, ectopic eruptions, supernumerary, or congenitally missing teeth.

Oral habits included severe bruxism resulting in generalized attrition of the primary teeth, especially the maxillary and mandibular anterior teeth. Moderate to severe wear facets also were present on the mandibular primary anterior teeth (Fig 4, next page).

All 20 primary teeth were present, caries free with sound enamel, and asymptomatic. The maxillary right and left central incisors exhibited Class II mobility apparently associated with the chronic digital sucking and bruxism.

A radiographic survey revealed that the patient's

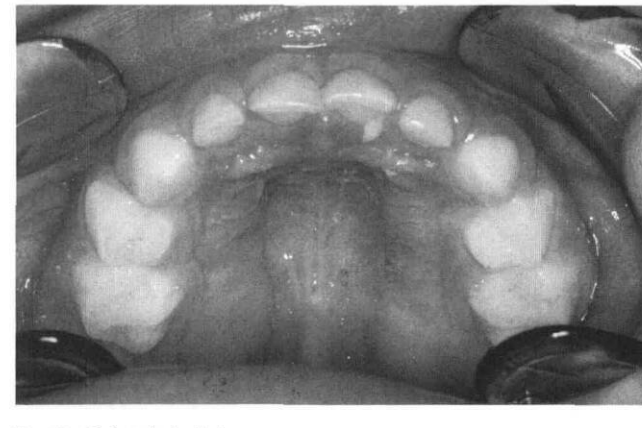

Fig 3. Palatal shelving.

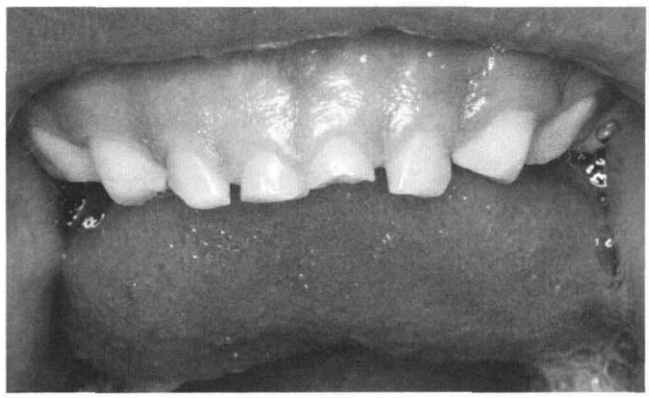

Fig 4. Severe attrition of the mandibular primary anterior teeth due to bruxism.

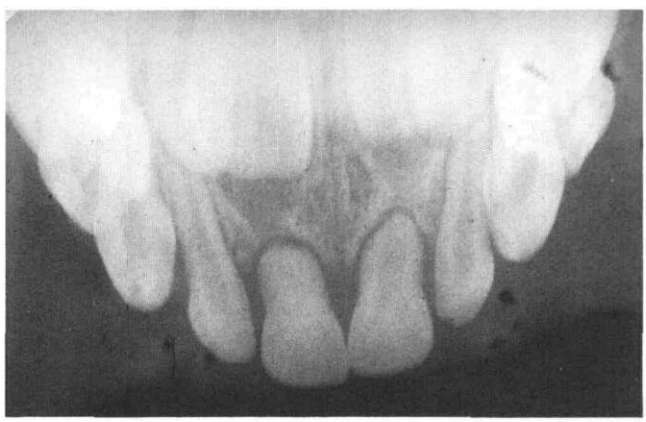

Fig 5. External root resorption, a widening of the periodontal ligament, and signs of calcific metamorphosis of the maxillary primary central incisors.

dental development was normal for a 4 year old. The maxillary primary central incisors exhibited external root resorption, a widened periodontal ligament, and signs of calcific metamorphosis (Fig 5).

The patient exhibited autistic-like behavior with periods of eye contact with the dentist. Her reaction to dental treatment was negative on the Frankl Scale (Frankl 1962). A Molt mouth prop was used during examination.

The patient exhibited poor oral-motor control, increased salivation, drooling, severe bruxism, and unique oral/digital hand movements between numerous episodes of stereotypic hand washing (Fig 6).

The patient's current management plan consists of placing her in a preschool developmental program offering speech, language, and occupational therapy. She will receive preventive and follow-up care by pediatric neurology, metabolic clinic, orthopedics, and pediatric dentistry.

Discussion

Fig 6. The autistic-like features of Rett syndrome with stereotypic hand washing and hand clasping.

This case illustrates the multidisciplinary medical findings for a 4-year, 3-month-old white female diagnosed as having Rett syndrome. The patient fulfills all diagnostic criteria of Rett syndrome as defined by Hagberg (1985) in Table 1. There seems to be a profound degeneration of early behavioral, social and psychomotor achievements, evolving communication dysfunction, dementia, loss of acquired purposeful hand skill, stereotypic hand washing, and the appearance of gait apraxia and truncal apraxia/ataxia. Before the child's fourth year of life, she had been diagnosed as having spastic encephalopathy, mild global developmental delay, cerebral palsy, and autism. The combination of diagnostic criteria (Hagberg 1985) simplifies the diagnosis of Rett syndrome. By applying the diagnostic criteria, Rett syndrome may be misdiagnosed less frequently (Budden 1986; Hagberg and Witt-Engerstrom 1986).

This case exemplifies some oral manifestations associated with Rett syndrome. Those evident in this case concur with the published medical literature. In addition, unreported oral findings were observed which include mild to severe generalized attrition due to bruxism, poor oral-motor control, palatal shelving, calcific metamorphosis of the maxillary left primary central incisor, and abnormal resorption and mobility of the maxillary primary central incisors. Many of these oral manifestations are not unique to Rett syndrome. It is likely that they may be related to trauma secondary to the excessive oral/digital habits, poor gait, and/or seizure activity. As more cases are studied it will become clear how consistently these oral manifestations occur. Presently, more attention should be paid to the unusual but distinctive oral/digital habits.

To date, no dental treatment recommendations have been reported. Patients with Rett syndrome have global developmental delay, autistic characteristics, severe to profound mental retardation, and loss of function in all domains. These medical findings may result in difficult patient management and dental treatment. Therefore, preventive dental treatment is essential. Long-term care should consist of close monitoring of bruxism, trauma,

increasing oral hygiene frequency with the help of parents, daily applications of topical fluoride gel or rinse, and more frequent preventive recall appointments.

Reding et al. (1966) reported bruxism in 15% of normal children and young adults. Budden (1986) and Holm (1986) reported an incidence of bruxism in patients with Rett syndrome of 85% and 43%, respectively. To decrease the severe problems associated with bruxism, Shafer et al. (1983) recommend the fabrication of nocturnal splints to immobilize or guide the movement of the mandible. McDonald et al. (1987) recommend the construction of a vinyl plastic bite guard to be worn at night to prevent the continuing abrasion of the teeth. Splint therapy may be contraindicated in patients with Rett syndrome due to the degree of mental retardation, poor oral-motor control, and excessive oral/digital habits. Iwata (1986) and Hanks (1986) found that immobilizing the hands interrupted hand-to-mouth habits and hand biting.

Prior to treating a Rett syndrome patient for comprehensive dental care with the use of sedation/general anesthesia, one should assess the degree of hypersalivation, apnea during wakefulness, severity of autism, and expected life span. Early treatment and establishing strict preventive guidelines for patients with Rett syndrome may obviate the necessity of sedation or general anesthesia.

Summary

Rett syndrome is a recently discovered, progressive neurological disorder that occurs exclusively in females early in life. Females with this rare and often misdiagnosed clinical entity have unusual but distinctive oral/digital habits. Dentists aware of Rett syndrome and its distinct manifestations can aid in early diagnosis and treatment of those afflicted with the disorder.

A report of a 4-year, 3-month-old white female with Rett syndrome are presented with a review of the literature. The characteristic features and diagnostic criteria of Rett syndrome are outlined. Previously reported oral/digital habits and manifestations of Rett syndrome are listed along with other unreported oral findings. Many of these oral manifestations are not unique to Rett syndrome. There is a likelihood that they may be related to trauma secondary to the excessive oral/digital habits, poor gait, and/or seizure activity.

This is believed to be the first report of Rett syndrome in the dental literature.

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Anesthetic lollipops

Lollipops containing the potent anesthetic fentanyl produced positive results with frightened or uncooperative children facing surgery.

Researchers at the University of Utah found using this form of premedication presented a pleasant, non-threatening method of achieving preoperative sedation prior to the use of general anesthesia in children.

The objective of using the anesthetic lollipop is to reduce or eliminate fear, hyperactivity, or even trauma frequently experienced by children as they leave the security of familiar surroundings and enter the unfamiliar and often frightening environment of the operating room.

In the study, the group of children who licked the fentanyl anesthetic lollipop had less activity and anxiety, and were more quickly anesthetized than childrenreceiving either no premedication or an oral sedative solution containing meperidine, diazepam, and atropine. Peak sedative effects occurred between 15 and 30 min after premedication for both the lollipop and oral solution groups.

Times to awake, response to verbal commands, and discharge from the recovery room were similar for all three groups. However, fewer of the patients receiving the lollipop required additional analgesics for control of pain postoperatively according to the researchers. There was no prolongation of postoperative recovery or increase in side effects in patients receiving the anesthetic lollipop.