Rubinstein-Taybi syndrome (RTS) is a human genetic disorder characterized by mental retardation and physical abnormalities including broad thumbs, big and broad toes, short stature, and craniofacial anomalies. The oral manifestations include small oral opening, pouting lower lip, retro/micrognathia, and higher arched, narrow palate. The purpose of this case report was to demonstrate the complicated dental treatment of a 12-year-old, developmentally disabled girl, living with a foster family, who suffered from RTS, extensive caries, and very poor oral hygiene. The patient demonstrated total lack of cooperation. The dental treatment had been carried out under general anesthesia (GA). Possible problems during GA in such patients are described. Fiberoptic video-assisted bronchoscope was prepared for the GA in case of airway emergency and/or difficult intubation. The GA process was uneventful, despite the extensive treatment delivered to the patient. Prospects for future good oral and dental status in this patient are questionable because of her extreme lack of cooperation.

KEYWORDS: RUBINSTEIN-TAYBI, DENTAL TREATMENT, ORAL MANIFESTATIONS, ANESTHESIA

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A 12-year-old, developmentally disabled girl was referred to the Department of Pediatric Dentistry at the Hebrew University—Hadassah School of Dental Medicine in Jerusalem, Israel, by her dentist for dental treatment. The patient was diagnosed as suffering from RTS. The patient was treated 4 years earlier under GA and needed additional dental treatment. She had all the characteristic features of RTS:

1. mental retardation;
2. broad thumbs (Figure 1);
3. big and broad toes;
4. short stature;
5. craniofacial anomalies like microcephaly, ptosis, beaked nose with low-lying philtrum, and epicanthic folds (Figure 2).

The patient lived with a foster family and attended a special school.

At the first visit, the patient was very friendly, but refused to be examined. She was rescheduled for examination under conscious sedation (diazepam and nitrous oxide). The patient did not fully cooperate, however, and only clinical examination could be performed.

The clinical examination revealed the following: (1) limited oral opening (20 mm); (2) micrognathia; (3) high, narrow-grooved palate; and (4) very narrow dental arches (Figure 3). The sequence and timing of eruption of the permanent dentition appeared normal. The patient had multiple malpositioned and crowded teeth, posterior cross bite, and anterior open bite. Severe gingivitis and extensive plaque and calculus accumulation were observed (Figure 4). Dental caries was evident on many teeth. A black stain could be observed on the posterior teeth.

Due to the extensive dental treatment, which required several visits, and the lack of cooperation, the patient was scheduled for dental treatment under GA.

Orthodontic consultation was made prior to the GA session, and minor treatment was recommended including extraction of premolars. The foster parents, however, refused any orthodontic treatment or, in particular, extractions of permanent teeth.

In the treatment session, 1.5 mg/kg of lidocaine 1%, followed by 1.5 mg/kg of propofol, were administered intravenously via 20 g cannula administered into the back of the patient's hand. Fiberoptic video-assisted bronchoscope was prepared in case of possible difficult airway management. Endotracheal intubation was then performed.

Anesthesia was maintained with isoflurane, carried by a mixture of air and oxygen. The dental treatment progressed uneventfully. Upon completion of the dental treatment (after 3 hours), the patient was extubated and transferred to the recovery room for further observation. Three hours later, she was released to the ward for overnight hospitalization. The patient was released from the hospital the next morning.

During GA, radiographs were taken (Figures 5 to 7), revealing multiple carious lesions—including the lower and upper incisors and impacted lower right second molar—and lack of eruption space of both second mandibular molars.
The treatment plan included:
1. a strict prevention protocol:
   a. cleaning the teeth;
   b. fluoride application;
   c. posttreatment meticulous oral hygiene instructions;
2. restorations of all carious teeth.

The posterior teeth were restored with amalgam, while the incisors were restored with composite resin for esthetic consideration.

One week after the treatment, neither pain nor sensitivity were reported. Oral hygiene instructions were given as well as dietary restrictions regarding reducing frequency of sugar consumption. A follow-up examination 6 months later—carried-out under lack of cooperation—disclosed intact restorations, but massive accumulation of plaque in the maxillary anterior region. The foster family could not imply proper oral hygiene. Topical application of Fluoride varnish (Duraphat 2.26% F ion, A. Nattermann & Cie.GmbH, Colonge, Germany) was performed, and the importance of maintaining oral hygiene was emphasized.

Discussion
This patient presented the typical manifestations of RTA. In addition, she lived with a foster family and did not maintain any oral hygiene. The foster family experienced extreme difficulties in providing oral hygiene and dental care due to her lack of cooperation. This, in turn, led to visiting a dentist only for emergencies.

The patient presented a high risk for caries due to cariogenic diet, lack of fluoride application, and poor dental health behavior. These factors, in addition to limited mouth opening, have led to treatment under GA, as recommended in previous reports.

In the present case, potential problems administering the endotracheal tube during GA were expected because of the limited mouth opening, the high-arched palate, and the micrognathia. In addition, the history of recurrent respiratory infections and the evidence for gastroesophageal reflux required extreme caution. Therefore, a fiberoptic video-assisted bronchoscope was prepared. The intubation and the anesthesia process were uneventful, and the fiberoptic video-assisted bronchoscope was not used.

In addition to the anesthesia-related risks, other risks in RTS patients have been reported. These include supraventricular and ventricular dysrhythmias, hypotonia, seizures, respiratory and cardiac failure, and delayed recovery from GA. In this case report, recovery from the GA was within the normal range (3 hours), and there were no signs of congenital heart disease—which has been reported in approximately one third of RTS patients. Thus, no prophylactic antibiotics were necessary.

Extraction of the crowded premolars, as suggested by the orthodontist prior to the dental treatment under GA, was not accepted by the foster parents, even though this may have improved the patient's look and malocclusion. The foster parents feared that the girl's behavior simply would not allow any orthodontic procedures.

Parental cooperation is a key factor in maintaining oral health in children with special needs who face difficulties in taking care of their own health.

In this case report, the authors hypothesized that, because of the radical and dramatic mode of dental treatment under general anesthesia, the foster parents would try harder to maintain the patient's oral hygiene to avoid future dental disease and subsequent treatment, as reported in previous studies. The girl's difficult behavior, however,
was apparently beyond the foster family's capability. Thus, despite the dental team's frustration, prospects for future good oral and dental status are questionable.

References