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# Anesthetic management of a child with Rubinstein-Taybi syndrome for cervical dermoid cyst excision

Füsun Bozkirli, Berrin Günaydin, Hülya Çelebi, and Didem Tuba Akçali

Gazi University Faculty of Medicine, Department of Anesthesiology, 06500 Ankara, Turkey

**Key words** Rubinstein-Taybi syndrome · Anesthesia

## Introduction

A limited number of cases of anesthetic management of Rubinstein-Taybi syndrome (RTS) has been reported since it was first diagnosed in 1963 [1]. The major features of this syndrome are short stature, mental retardation, antimongoloid slant of the eyes, beaked nose, hypoplastic maxilla, broad thumbs and toes, congenital heart defects, and microdeletion of chromosome 16p13. Features such as hypoplastic maxilla, narrow palate, micrognathia, and small opening of the mouth may play an important role in endotracheal intubation [2–4]. We present our anesthetic management of a 5-year-old child undergoing operation twice for a neck mass.

## Case report

The patient was diagnosed with RTS when he was 1 month old. He was born at term by normal spontaneous delivery. His birth weight and length were 3050 g and 50 cm, respectively. He had recurrent respiratory infections due to regurgitation, which might have been caused by the immaturity of the esophageal muscles. On admission to our hospital, this boy had mental and motor retardation, bilateral clinodactyly and broad thumbs, pectus excavatum, and micrognathia, all of which are characteristics of this syndrome (Fig. 1). Thyroid ultrasonography showed a multilocular cystic $33 \times 20$ mm mass anterior to the thyroid gland. There was no mass activity on thyroid scintigraphy. The total blood count and other laboratory test results were within normal ranges. No gastroesophageal reflux was observed on esophagogastrography. The right testis had not descended, and the left testis was retractile. The results of cranial computerized tomography and chromosomal analysis were normal. He had no cardiac defects. He was diagnosed as having the autosomal dominant mutagenic form of RTS.

There was an uneventful history of operation and anesthesia for a midline neck mass diagnosed as a thyroglossal cyst that was performed at another center 1 year previously. On this occasion, the midline neck mass was above the previous one and was thought to be a recurrent thyroglossal cyst preoperatively. The patient's height was 90 cm (<3 percentile), and his weight was 14 kg (3–10 percentiles). He fasted for 8 h and was not premedicated before induction of anesthesia. His heart rate was 130 beats·min$^{-1}$ and his $SpO_2$ was 99%. After preoxygenation, anesthesia was induced by 50% $N_2O/O_2$ mixture in sevoflurane via a face mask. After intravenous access had been obtained, 0.5 mg kg$^{-1}$ of atracurium was injected, and endotracheal intubation was performed without difficulty with an uncuffed tube that had an internal diameter of 4.5 mm. Anesthesia was maintained with 50% $N_2O/O_2$ mixture in sevoflurane. The neck mass was excised. At the end of anesthesia, spontaneous ventilation was promptly reestablished. After reversal of residual muscle paralysis with 0.5 mg of neostigmine and 0.25 mg of atropine, the endotracheal tube was removed. Rectal paracetamol was administered for postoperative pain. No significant problem was encountered during induction, maintenance, and extubation. The pathologic diagnosis of the mass was dermoid cyst.

*Address correspondence to*: B. Günaydin, Kızılırmak mahallesi 8. sokak 7/35, C-Blok Balgat 06530, Ankara, Turkey
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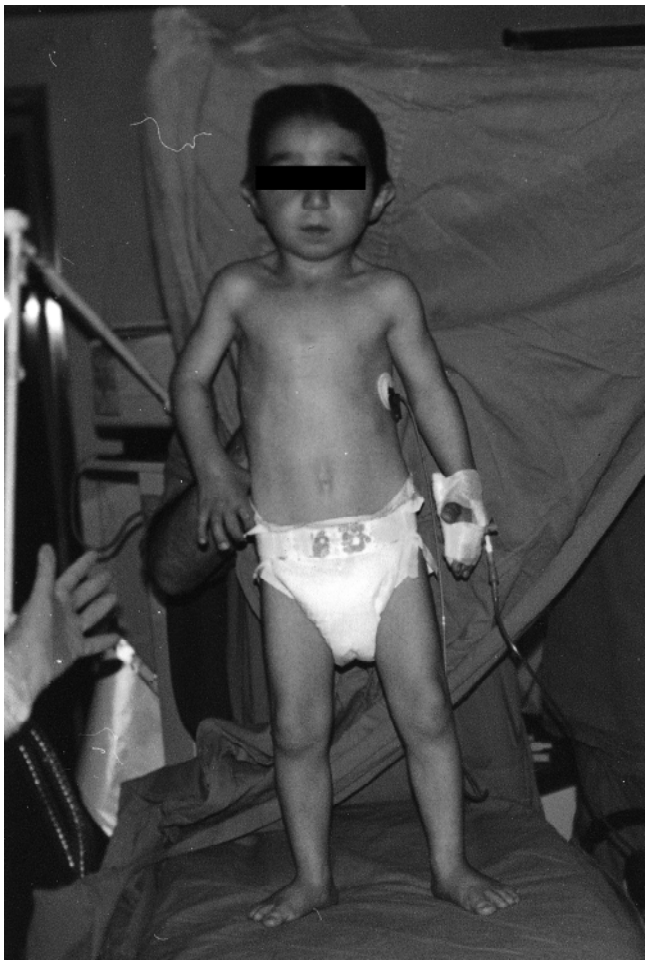

**Fig. 1.** Physical appearance of a child with Rubinstein-Taybi syndrome

## Discussion

The majority of neck masses requiring surgical therapy in children are of congenital origin. Dermoid cysts in the midline and paramedian neck are often misdiagnosed as thyroglossal duct cysts [5]. In a reported case of a thyroglossal duct cyst associated with a dermoid cyst, two cysts were not attached, but the presence of two lesions was further evidence for a possible developmental anomaly [6].

The facial features associated with RTS could cause difficulties in intubation. In our patient the length of the neck was not short, the Mallampati score was 1, and the other facial features did not suggest difficult intubation. This is one of the reasons we used atracurium. The other reason is that there have been reports of arrhythmias due to succinylcholine administration in RTS [4,7]. Although Critchley and co-workers [8] reported no arrhythmias due to succinylcholine, Stirt [4] anesthetized a child with RTS three times, and severe arrhythmias occurred after the administration of succinylcholine. We did not encounter any difficulties during intubation.

Although the duration of the operation was short enough to allow the use of a laryngeal mask airway (LMA), we did not use it because an immobile LMA was not suitable for neck surgery and there was a risk of aspiration of the gastric contents.

We avoided using halothane because it can sensitize the myocardium to the dysrhythmic effects of catecholamines and cause dysrhythmias, such as nodal rhythm and ventricular extrasystoles in case of hypocapnia [9]. We maintained anesthesia by an $O_2/N_20$ mixture in sevoflurane.

Critchley et al. [8] performed general anesthesia combined with caudal block and provided postoperative pain relief by caudal anesthesia because their operation involved the inguinal region. In our patient we used rectal paracetamol for postoperative analgesia. Intravenous opioid infusion would also be a choice after very painful operations.

In summary, we have presented our anesthetic management of a 5-year-old boy with RTS who underwent surgery for a cervical mass in which endotracheal intubation was performed uneventfully. RTS carries important risks for anesthesia because of potential difficulty in intubation and the necessity to use short-acting depolarizing muscle relaxants that may lead to cardiac arrhythmias. When all precautions are taken against difficult intubation and other adverse events, anesthesia can be performed successfully.

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