Images in Anesthesia: Congenital tracheal stenosis in a boy with Rubinstein-Taybi syndrome

FIGURE Three-dimensional reconstruction of the chest computed tomography scan, with virtual endoscopy images at different levels, showing the morphologic characteristics of the tracheal stenosis. In its narrowest portion, the diameter of the stenosis was 3.0 mm.

RUBINSTEIN-TAYBI syndrome (RTS) is a complex entity, characterized by facial changes, broad thumbs and great toes, postnatal growth retardation, cognitive defects and multiple malformations.¹ No airway abnormalities are described in RTS other than post-cricoid web² and tracheoesophageal compression due to vascular ring.³ However, because craniofacial changes often include micrognathia, problems with intubation during anesthesia may occur with great frequency.^{4,5} We report the first case of congenital tracheal stenosis in RTS.

A 46-month-old Caucasian male, diagnosed soon after birth as being affected by RTS, was admitted for surgical correction of bilateral cryptorchidism. At

admission, the characteristic face and the typical thumb and toe changes were observed. Laparoscopy followed by orchiopexy was planned, and because of the presence of a high arched palate and marked hypoplastic maxilla, difficulties in intubation were expected. While no problems in passing the vocal cord with the tracheal tube were encountered, an unpredictable resistance thereafter was found. Fibreoptic bronchoscopy was performed and a severe concentric tracheal stenosis with complete cartilaginous O-rings was detected, whose characteristics were better defined by spiral computed tomography scan three-dimensional reconstruction (Figure). No vascular rings or other great vessel abnormalities were detected.

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