

## Growth causes difficult tracheal intubation in a patient with Kniest dysplasia

YOSHIO SEGAWA, HARUO ENOMOTO, TAKAKO NAKAGAWA, and YASUSHI IJIMA

Department of Anesthesia, NTT West Hospital, 2-6-40 Karasugatsuji, Tennouji-ku, Osaka 543-8922, Japan

**Key words** Kniest dysplasia · Difficult tracheal intubation · Growth

### Introduction

Kniest dysplasia is a severe chondrodysplasia caused by the defective formation of type II collagen and characterized by short trunk and limbs, kyphoscoliosis, midface hypoplasia, severe myopia, and hearing loss [1,2]. We describe a case in which we encountered difficult tracheal intubation after growth at the fourth general anesthesia, although the preceding three anesthetic managements were quite uneventful.

### Case report

A 11-year-old girl (weight 21.5 kg, height 106.5 cm) was scheduled for ophthalmic surgery. She had the following features (shown in Fig. 1 at the age of 6 years): short trunk and limbs, relatively large head, midface hypoplasia, tight curvature of thoracic and lumbar spine with wedge-shaped vertebrae, inability to flex the fingers, limitation of shoulder movement, enlargement of elbow and knee with a distinctive dumbbell appearance of the long bones, and hearing loss. There had been no anesthetic problem in the past three ophthalmic surgeries at the age of 6 years (twice at a weight of 16.0 kg and a height of 87.7 cm, and once at a weight of 17.0 kg and a height of 90.0 cm) [3]. We induced anesthesia with 6% sevoflurane by mask because venous access was very difficult. After venous cannulation, we achieved muscle relaxation with vecuronium bromide 4 mg iv. Mask ven-

tilation was easy. Laryngoscopy was carried out as usual with a Macintosh blade no. 2; however, we could only see the tip of the epiglottis and could not visualize the vocal cords. Intubation was very difficult. Light pressure was applied on the larynx, and we kept the 4.5-mm cuffed spiral tube with stylet well anterior, up against the epiglottis. Intubation was successful at the fourth trial. After intubation, the anesthetic course was uneventful. The patient was extubated in the operating room. After extubation, there was no stridor at the glottis, and she went back to the ophthalmic ward.

### Discussion

We can find no report of anesthesia of a patient with Kniest dysplasia. Our patient was born with hearing loss. She has cataract, retinal detachment, and glaucoma [3,4]. Therefore, she needed four operations. We had no problem in anesthetic management at age 6. We encountered no difficult tracheal intubation in the previous three anesthetics. This time, after laryngoscopy, we could only see the tip of the epiglottis. It was classified as grade 3 according to Cormack and Lehane [5], so that intubation had to be blind. We consider that anatomical changes caused the difficult tracheal intubation. The shape of the larynx gradually becomes anatomically the same as that of an adult around 8 years of age. The difference is the size. The larynx is positioned at the fourth vertebra at birth. It goes down to the upper end of the fifth vertebra at the age of 6 or 7 years, and between the fifth and the sixth vertebrae in the adult [6,7]. The patient with Kniest dysplasia also has midface hypoplasia. These would make visualization of the glottis impossible at age 11. Fortunately, mask ventilation was easy. We tried blind intubation several times. Finally, we successfully intubated her.

In conclusion, we have reported our experience of a patient with Kniest dysplasia. We encountered difficult



**Fig. 1.** Photograph of the patient at the age of 6 years

endotracheal intubation, but not in the previous three anesthetics. Growth might have caused this difficult tracheal intubation.

### References

1. Wilkin DJ, Artz AS, South S, Lachman RS, Rimoin DL, Wilcox WR, McKusick VA, Stratakis CA, Francomano CA, Cohn DH (1999) Small deletions in the type II collagen triple helix produce Kniest dysplasia. *Am J Med Genet* 16:105–112
2. Spranger J, Winterpacht A, Zabel B (1997) Kniest dysplasia: Dr. W. Kniest, his patient, the molecular defect. *Am J Med Genet* 69:79–84
3. Makino I, Watanabe M, Okamoto N, Ohguro N, Fukuda M, Emi K (1997) A case of retinal detachment in Kniest dysplasia treated with vitreous surgery. *Nippon Ganka Gakkai Zasshi (J Jpn Ophthalmol Soc)* 101:734–737
4. Kagotani Y, Takao K, Nomura K, Okubo K (1995) Two cases of Kniest dysplasia—ocular manifestations. *Nippon Ganka Gakkai Zasshi* 99:376–383
5. Cormack RS, Lehane J (1984) Difficult tracheal intubation in obstetrics. *Anaesthesia* 39:1105–1111
6. Eckenhoff JE (1951) Some anatomic considerations for the infant larynx influencing endotracheal anesthesia. *Anesthesiology* 12: 401–410
7. Okuaki A (1995) Endotracheal intubation. In: Inada Y (ed) *Saishinmasuikagaku* (in Japanese). Kasseidoushuppan, pp 747