J Anesth (2001) 15:53–56

Journal of
Anesthesia

Anesthetic management of a patient with Klippel-Feil syndrome

Sukanya Mitra, K.K. Gombar, Kalpana Sharma, Charu Deva, and Arjun Das

Department of Anesthesia and ENT, Government Medical College, Sector 32, Chandigarh 160047, India

Key words Klippel-Feil syndrome · Cleft palate · Anesthesia · Complications

Introduction

Klippel-Feil syndrome, first described in 1912 [1], is characterized by shortness of the neck resulting from reduction in the number of cervical vertebrae. The classic triad of short neck, low posterior hairline, and limited neck movements is present in 50% of cases. The syndrome is often associated with congenital anomalies of other parts of the skeletal system [1,2], along with congenital cardiac and genitourinary defects [2,3]. These patients have potential cervical instability, are at high risk of neurological damage during laryngoscopy, intubation and positioning [4], and thus pose a great challenge to the anesthetist. We report a patient with Klippel-Feil syndrome who underwent repair of cleft palate, and we discuss anesthetic considerations in management.

Case report

A 12-year-old boy weighing 25 kg was referred to our hospital for the repair of cleft palate. There was no definitive history of obstructive sleep apnea. The child had low posterior hairline, short neck, and right sided torticollis. On examination he had a repaired scar of a cleft lip, grade III cleft palate (Fig. 1), and deviated nasal septum with malocclusion of teeth. The thumb of his right hand was absent, and he had Sprengel's deformity (elevated and small scapula) (Fig. 2). He had restricted neck movements with minimal extension at the cervical vertebrae. He had fusion of the cervical vertebrae at the level of C2–3 and C5–6. The patient had a severe degree of kyphoscoliosis (Cobb angle was 40°) and could only lie down with two pillows below his occiput. On auscultation, he had minimally decreased air entry of the right lung. His higher function examination suggested poor mentation, and this was further corroborated by IQ testing, which revealed an IQ rating of 73. He had no muscle weakness or other neurological symptoms. His cardiovascular system was unaffected, with normal two-dimensional echocardiography. Ultrasound echography of his abdomen showed absence of the right kidney. He was unable to perform pulmonary function tests because of his cleft palate. However, hematological, coagulation, and biochemical investigations were unremarkable. His chest X-ray did not reveal any significant changes.

The preoperative evaluation of airway (temporomandibular joint function) revealed a mouth opening of more than 40 mm. Mallampati classification was not possible because of the associated cleft palate. His extension of head was more than two-thirds restricted. The patient had dental malocclusion. However, radiological examination for estimation of mandibular space was not done.

After informed consent from his parents, the child was premedicated with 75 mg ranitidine orally on the night before and on the morning of surgery and received 0.1 mg glycopyrrolate intramuscularly 1 h before operation.

In the operating theater, he was connected to a multichannel monitor (Datex-AS-3 Light, Helsinki, Finland), and his heart rate, noninvasively measured blood pressure, electrocardiogram, arterial oxygen saturation (SpO_2), and end-tidal carbon dioxide ($ETCO_2$) were

Address correspondence to: S. Mitra c/o Dr. D. Basu, Dept. of Psychiatry, PGIMER, Chandigarh 160012, India
Received: December 13, 1999 / Accepted: August 11, 2000

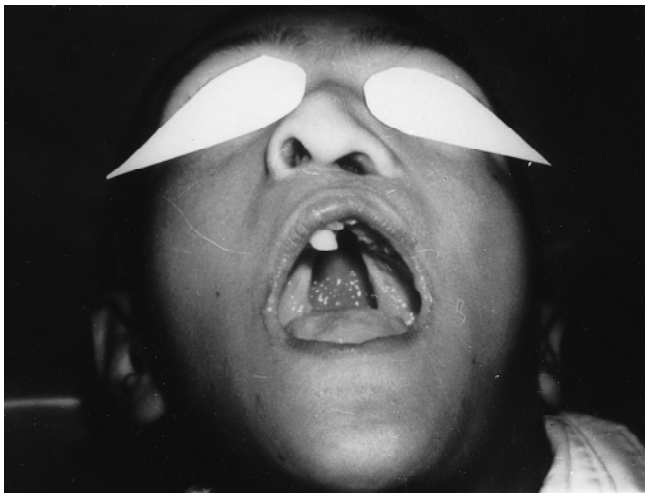

Fig. 1. Photograph of the oral cavity of the patient

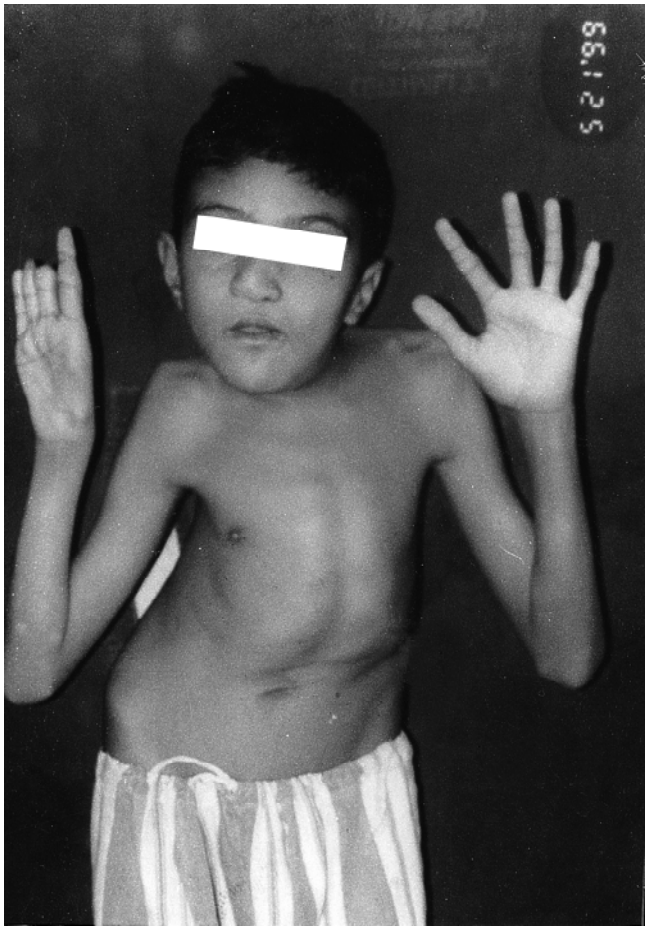

Fig. 2. Photograph of the patient with Klippel-Feil syndrome

continuously monitored. The patient was made to lie down supine with two pillows below his occiput. He could lie down flat only with his neck turned towards the right side. An intravenous line was started on the dorsum of the hand, and 5 mg metoclopramide was administered intravenously. Because of the anticipated difficult intubation, an inhalational induction was planned. Anesthesia was induced with 50% nitrous oxide in oxygen with incremental doses of sevoflurane with a maximum concentration of 5% using a Mark V (Ohmeda, Hatfield, UK) vaporizer. When an adequate depth of anesthesia had been achieved, gentle direct laryngoscopy was performed to assess the airway. Once the airway was assessed, 1.5 mg · kg^{-1} suxamethonium was given and the trachea was intubated with a 5.5-mm cuffed endotracheal tube (Portex, Kent, UK). Bilateral equal air entry was confirmed and the tube position was secured.

The patient was then paralyzed by 10 mg atracurium, and 1 mg morphine was given. Intermittent positive pressure ventilation was started with 66% N_2O in oxygen with 0.4%–0.6% isoflurane using Jackson-Ree's modification of Ayre's T piece. Surgery lasted for 2 h 30 min. During this period, the blood loss was minimal and the child received no transfusion.

Maintenance of anesthesia was uneventful, and at the end of the procedure the residual neuromuscular blockade was reversed by 0.05 mg·kg^{-1} neostigmine and 0.02 mg·kg^{-1} atropine. The endotracheal tube was removed in the postanesthesia care unit after 4 h. He had no adverse neurological sequelae. The child was shifted to the recovery room breathing 35% oxygen. He was subsequently shifted to the intensive care unit (ICU) for observation and then transferred to the ENT ward from the ICU the next morning.

Discussion

Patients with Klippel-Feil syndrome pose numerous problems to the anesthetist. Some of these are due to the essential features of the syndrome, i.e., short neck and absence or fusion of some of the cervical vertebrae, along with anomalies in the adjoining cranial area, thus limiting neck movements as well as compromising cervical stability [4]. Hence, there is an obvious anesthetic risk of spinal cord injury during laryngoscopy, intubation, and positioning of the patient [5]. Associated abnormalities at the atlanto-occipital junction, spinal canal stenosis, or scoliosis also increases the risk of neurological damage during or following anesthetic procedures, essentially due to mechanical factors such as pressure, torsion, or rotation of the neck [2,6]. Furthermore, sudden rotatory movements of the neck in these patients may precipitate syncopal attacks, and hence anesthetists should be specially cautious to avoid forceful neck movements when managing such patients [5].

In a patient with anticipated difficult intubation, awake intubation would have been a method of choice.

However, our patient was a child with a low IQ who became very upset and refused to cooperate in such an attempt. Pediatric patients, unlike adults, almost always require general anesthesia [7]. In fact, they become severely stressed by attempts at awake intubation [8]. Hence, the prospect of performing awake intubation was abandoned at the outset. The use of a reinforced laryngeal mask airway in cleft palate surgery has been found a suitable alternative to the endotracheal tube, but its use may be associated with airway obstruction and displacement [9]. Fiberoptic-guided intubation is probably the method of choice in these patients [10]. However, the nonavailability of fiberoptic equipment prompted us to perform direct laryngoscopy with adequate precautions. An inhalational induction was performed using incremental concentrations of sevoflurane for a patent airway. The impressive lack of airway irritation with low blood gas coefficient makes sevoflurane useful for induction of anesthesia, especially in children [11]. Once an adequate depth of anesthesia was achieved, a direct careful laryngoscopy was performed. Although the trachea could be intubated by using a direct laryngoscope, we had orpharyngeal airway, laryngeal mask airway, and percutaneous needle tracheostomy sets in our anesthesia trolley to combat any emergency situation.

Klippel-Feil syndrome is noted for the large number of associated congenital anomalies. For the anesthetist, it is essential to be aware of the ones that can be relevant for anesthetic management.

First, skeletal anomalies represent a common feature of this syndrome and include, among many others, a 60% prevalence of significant scoliosis. Sprengel's deformity has been identified in 25%–35% of these patients [2,12]. Our patient had both conditions. These may compromise the ventilatory capacity and make patient positioning more difficult. The presence of severe kyphoscoliosis with surgery in the oral cavity makes these patients particularly vulnerable to hypoxemia.

Second, cardiovascular congenital anomalies have been recognized in these patients, with a prevalence of 4.2%–14% [12,13]. Several lesions may occur, the commonest being ventricular septal defects, but also patent ductus arteriosus [12,14]. These patients may further develop cardiac failure preoperatively. Other lesions reported are mitral valve prolapse, bicuspid aortic valve, and coarctation of the aorta [13]. Preoperative evaluation to rule out such conditions should be mandatory. Fortunately, our patient did not have any such defects.

Third, other visceral congenital anomalies, especially genitourinary anomalies, may also occur in patients with Klippel-Feil syndrome and these may be of relevance in anesthetic management. Genitourinary anomalies have been identified in 64% of these patients [3]. Our patient had an absent right kidney, but renal function test results were normal.

The presence of other anomalies, such as cleft palate, mandibular malformations, and micrognathia, in association with Klippel-Feil syndrome may make airway management and oxygenation difficult [15]. Our patient had a large cleft plate along with scoliosis, thus compounding the problem of maintaining ventilation in the perioperative period.

Finally, our patient had a low IQ (73), suggesting borderline intelligence. Low intelligence may give rise to difficulty in communication and comprehension in the perioperative period. The problem may be further complicated if the patient happens to have other anomalies restricting his sensory functions, such as deafness and other ocular anomalies, which have indeed been described in such patients [16,17].

Careful preoperative evaluation to exclude associated congenital anomalies, avoidance of manipulation during laryngoscopy, intubation, and positioning, and better understanding of associated pathophysiology may improve the outcome in a patient with Klippel-Feil syndrome.

References

1. Klippel M, Feil A (1912) Un cas d' absence des vertebres cervicals. Soc Anat Paris Bull Mem 14:185
2. Hensinger RN, Lang JE, MacEwen GD (1974) Klippel-Feil syndrome. A constellation of associated anomalies. J Bone Joint Surg (Am) 56:1246–1253
3. Moore WB, Mathews TJ, Rabinowitz R (1975) Genitourinary anomalies associated with Klippel-Feil syndrome. J Bone Joint Surg (Am) 57:355–57
4. Nagib MG, Maxwell RE, Chou SN (1984) Identification and management of high—risk patients with Klippel-Feil syndrome. J Neurosurg 61:523–530
5. Naguib M, Farag H, Ibrahim AEW (1986) Anaesthetic considerations in Klippel-Feil syndrome. Can Anaesth Soc J 33:66–70
6. Gunerson H, Greenspan RH, Glaser H (1967) The Klippel-Feil syndrome: genetic and clinical reevaluation of cervical fusion. Medicine 46:491–512
7. Steward DJ (1999) Management of the difficult pediatric airway. Anesth Analg Suppl (March):93–97
8. Cook-Sather SD, Tulloch HV, Subina ML, Nicholson SC, Schreiner MS (1995) A comparison of awake versus paralysed tracheal intubation in infants for pyloromyotomy. Anesthesiology 83:A1151
9. Gombar KK, Gombar S (1998) Use of reinforced laryngeal mask airway (RLMA) in children undergoing cleft palate surgery Anaesthesiologie Intensivmedizen Notfall Medizin-Schmerz Therapie. Supplement 33:428
10. Daum RE. Jones DJ (1988) Fibreoptic intubation in Kilppel-Feil syndrome. Anaesthesia 43:18–21
11. Naito Y, Tamai S, Slunga K, Fujimori R, Mori K (1991) Comparison between sevoflurane and halothane for paediatric ambulatory anaesthesia. Br J Anaesth 67:387–389
12. Thomsen MN, Weber M, Niethard FU (1997) Scoliosis and congenital anomalies with Klippel-Feil types I–III. Spine 22:396–401
13. Morrison SG, Perry LW, Scatt LP III (1968) Congenital brevicollis (Klippel-Feil syndrome) and cardiovascular anomalies. Am J Dis Child 115:614–620

14. Falk PH, Mackinnon J (1976) Klippel-Feil syndrome associated with aortic coarctation. Br Heart J 38:1220–1221
15. Widgerow AD (1990) Klippel-Feil anomaly, cleft palate, and bifid tongue. Ann Plast Surg 25:216–222
16. Palant DI, Carter BL (1973) Klippel-Feil syndrome and deafness. A study with polytomography. Am J Dis Child 123:218–221
17. Kirkham TH (1970) Duane's retraction syndrome and cleft palate. Am J Ophthalmol 70:209–212