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## CLINICAL INFORMATION

# Airway management in newborn with Klippel–Feil syndrome



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### KEYWORDS

Neonatal;  
Difficult airway;  
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Congenital heart disease;  
Surgery;  
Critical care

**Abstract** Klippel–Feil syndrome (KFS) has a classical triad that includes short neck, low hair line and restriction in neck motion and is among one of the congenital causes of difficult airway. Herein, we present a 26-day, 3300 g newborn with KFS who was planned to be operated for correction of an intestinal obstruction. She had features of severe KFS. Anesthesia was induced by inhalation of sevoflurane 2–3% in percentage 100 oxygen. Sevoflurane inhalation was stopped after 2 min. Her Cormack Lehane score was 2 and oral intubation was performed with 3.5 mm ID non-cuffed endotracheal tube in first attempt. Operation lasted for 45 min. Following uneventful surgery, she was not extubated and was transferred to the newborn reanimation unit. On the postoperative third day, the patient died due to hyperdynamic heart failure. This case is the youngest child with Klippel–Feil syndrome in literature and on whom oral intubation was performed. We also think that positioning of this younger age group might be easier than older age groups due to incomplete ossification process.

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### PALAVRAS-CHAVE

Neonatal;  
Vias aéreas difícil;  
Anormalidades congênitas;  
Doença cardíaca congênita;  
Cirurgia geral;  
Cuidados intensivos

### Manejo de vias aéreas em recém-nascido com síndrome de Klippel-Feil

**Resumo** A síndrome de Klippel-Feil (SKF) envolve uma tríade clássica que inclui pescoço curto, linha de implantação dos cabelos baixa e restrição do movimento do pescoço e é uma das causas congênitas de via aérea difícil. Apresentamos o caso de uma recém-nascida, de 26 dias, 3.300 g de peso, com SKF, que foi agendada para cirurgia de correção de uma obstrução intestinal. A paciente apresentava características acentuadas da SKF. A anestesia foi induzida com inalação de sevoflurano a 2-3% em 100% de oxigênio. A inalação de sevoflurano foi interrompida após dois minutos. O escore de Cormack-Lehane da paciente era 2 e a intubação orotraqueal foi feita na primeira tentativa, com tubo endotraqueal de 3,5 mm ID sem balão. O tempo de cirurgia foi

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de 45 minutos. Após a cirurgia sem intercorrências, a paciente foi transferida para a unidade de reanimação neonatal ainda intubada. No terceiro dia de pós-operatório, foi o óbito por causa de uma insuficiência cardíaca hiperdinâmica. Esse caso apresenta a criança mais jovem com síndrome de Klippel-Feil na literatura e na qual a intubação orotraqueal foi feita. Acreditamos também que o posicionamento de pacientes dessa faixa etária mais jovem pode ser mais fácil do que o de pacientes de faixas etárias mais altas, por causa do processo de ossificação incompleto. © 2014 Sociedade Brasileira de Anestesiologia. Publicado por Elsevier Editora Ltda. Este é um artigo Open Access sob uma licença CC BY-NC-ND (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## Introduction

Klippel-Feil syndrome (KFS) has an incidence of 1:42,000 births and is among one of the congenital causes of difficult airway. The classical triad includes short neck, low hair line, restriction in neck motion, and fusion of at least two cervical segments. Short neck with a limited range of movement and cervical instability can lead to neurological damage during laryngoscopy, intubation and positioning for surgery.<sup>1</sup>

## Case description

Herein, we present a 26-day, 3300 g newborn with KFS who was planned to be operated for correction of an intestinal obstruction. On physical examination, she had features of severe KFS: including short neck, restriction in neck motion, high palate, class II modified Mallampati score, low hair line and atypical facial appearance (Fig. 1). The echocardiogram revealed ASD, VSD, aortic pseudocoarctation, PDA, wide coronary sinus, persistent pulmonary HT. Blood pressure, oxygen saturation, and ECG were monitored in the operating room. Various large blades of Miller and Macintosh type, laryngeal mask airways, tracheal tubes with stilettes, fiberoptic bronchoscope, and a tracheostomy set were kept ready. Anesthesia was induced by inhalation of sevoflurane 2–3% in percentage 100 oxygen. Sevoflurane inhalation was stopped after 2 min. Her Cormack Lehane score was 2 and oral intubation was performed with 3.5 mm ID non-cuffed



**Figure 1** Low hair line and atypical facial appearance of the patient.

endotracheal tube in the first attempt. Correction of ET tube placement was done with auscultation and capnography. Anesthesia was maintained with 60% air in oxygen with 1–1.5% sevoflurane and Duodenoduodenostomy was done for the partial obstruction in duodenum. Operation lasted for 45 min. Following uneventful surgery, she was not extubated and was transferred to the newborn reanimation unit. On the postoperative third day, the patient died due to hyperdynamic heart failure.

## Discussion

Klippel-Feil syndrome is a rare disease, initially reported in 1912 by Maurice Klippel and André Feil from France, characterized by the congenital fusion of any 2 of the 7 cervical vertebrae. The classical triad includes short neck, low hair line, restriction in neck motion, and fusion of at least two cervical segments. Patients with Klippel-Feil syndrome usually present with the disease during childhood but this may also present later in life.<sup>2,3</sup> Our case was detected in the neonatal period and has classical triad signs of the syndrome. Difficult airway is the main problem of this group of patient in anesthesiology and intensive care medicine. In literature, most of the KFS cases involve adult age groups and is associated with difficult airway problems.<sup>1,2</sup> Stallmer et al. have recently published the management of 10 pediatric patients with KFS concluding that the airway of these patients can be settled without difficulty in a high percentage of cases similar to our case.<sup>4</sup> The youngest case in the literature is two-month-old girl. To the best of our knowledge, this case is the youngest one in the literature.

In that case, in order to avoid traumatization, LMA (size 1) was inserted with the reverse technique. This case is the youngest child with Klippel-Feil syndrome and on whom oral intubation was performed. We also think that positioning of this younger age group might be easier than older age groups due to incomplete ossification process. More extensive fusions tend to be associated with other defects involving the cardiovascular, respiratory, and genitourinary systems. Cardiovascular anomalies occur in 15–30% of the cases. The most common problem is the ventricular septal defect.<sup>3</sup> Heart failure is important in postoperative care process. Our case has ASD, VSD, aortic pseudocoarctation, wide coronary sinus and persistent pulmonary HT. The syndrome also may be associated with abnormalities of the head and face, skeleton, sex organs, muscles, brain and spinal cord, arms, legs, fingers and heart defects. In literature the only

KFS case with GI system anomalies was colon duplication.<sup>5</sup> Upper GI system abnormalities might cause nutritional disorders and respiratory problems similar to our case. Detailed preanesthetic evaluation of organ systems especially including airway and cardiovascular system is crucial in KFS.

### Conflicts of interest

The authors declare no conflicts of interest.

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