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## Clinical Reports

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### Anaesthetic considerations in Klippel-Feil syndrome

Mohamed Naguib MB BCH MSc,  
Hesham Farag MB BCH FFARCS,  
Abd El Wahab Ibrahim MB BCH FRCS

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*Anaesthesia for the repair of a large occipito cervical encephalomyelocele in a neonate with Klippel-Feil syndrome is described. The fusion of the cervical spines, a short neck, low posterior hair line and Sprengel's deformity, which were present in this patient, collectively indicated Klippel-Feil syndrome. In addition to the usual stigmata of the syndrome, this patient had a large encephalomyelocele and persistent patent ductus arteriosus complicated by congestive heart failure. Patients with this syndrome are vulnerable to cervical spinal cord injury and are at high risk for neurological injury not only during laryngoscopy and intubation but thereafter. Implications of Klippel-Feil syndrome for the anaesthetist are reviewed and discussed.*

Klippel-Feil syndrome, first described in 1912,<sup>1</sup> is characterized by shortness of the neck resulting from reduction in the number of cervical vertebrae or the fusion of several vertebrae into an osseous mass. The posterior hair line is low and the movement of the neck is limited.<sup>1,2</sup> The syndrome is often associated with congenital anomalies of other skeletal parts of the same segments, such as

Sprengel's deformity. Visceral congenital anomalies associated with the syndrome such as cardiovascular and genito-urinary anomalies, may also be present.<sup>2</sup>

Although much has been written in the surgical literature about this syndrome, no review of the anaesthetic problems associated with the condition has been published.

#### Case Report

A full-term three-week-old boy weighing 3 kg was referred to the hospital for repair of a craniocervical encephalomyelocele. The baby was delivered by Caesarean section and his Apgar Scores were seven and eight after one and five minutes respectively. The mother was 45 years old and was gravida 12, para 12, and the father was 65 years old. Both parents were healthy and there was no family history of medical disorders. The baby had hydrocephalus with a large occipito cervical encephalomyelocele, a short webbed neck with limited range of movement of the neck in the lateral direction, low posterior hair line, Sprengel's deformity (both scapulas were elevated and small) (Figure 1) and penile hypospadias. A patent ductus arteriosus, complicated by congestive heart failure was also detected. Roentgenograms and computerized tomography (CT scan) showed fusion of the C<sub>2-3</sub> vertebral bodies with spine bifida in all cervical and upper six dorsal vertebrae (Figures 2 and 3). In addition a CT scan disclosed a large dilatation of the ventricular system with communication of the encephalocele with the ventricular cavity. A diagnosis of Klippel-Feil syndrome was made. Congenital rubella syndrome was excluded by the absence of rubella-specific IgM (Rubazyme). Total IgM

#### Key words

ANAESTHESIA: paediatric; COMPLICATIONS: Klippel-Feil syndrome; GENETIC FACTORS: congenital defects.

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From the Departments of Anaesthesiology and Neurosurgery, King Faisal University, King Fahd Hospital, P.O. Box 2208, Al-Khobar, Saudi Arabia, 31952.

Address correspondence to: Dr. M. Naguib.



FIGURE 1 Photograph of right side of the head showing shortness of the neck and encephalomyelocle of the craniocervical region. The parietal region had been shaved in preparation for IV cannulation.

level was reported to be  $113 \text{ mg} \cdot \text{dl}^{-1}$  (normal =  $27\text{--}139 \text{ mg} \cdot \text{dl}^{-1}$ ).

Heart failure was controlled with digoxin  $0.02 \text{ mg}$  given twice daily, furosemide and a potassium supplement. The preoperative digoxin level was  $1.5 \text{ ng} \cdot \text{ml}^{-1}$  (therapeutic range  $0.9\text{--}2.0 \text{ ng} \cdot \text{ml}^{-1}$ ).

A preoperative electrocardiogram showed biventricular hypertrophy and axis deviation of  $+30^\circ$ . A chest x-ray disclosed a prominent aorta and pulmonary artery and cardiac enlargement. Haemoglobin was  $10.1 \text{ g} \cdot \text{dl}^{-1}$  and blood urea and serum electrolytes were within the normal range.

The patient was premedicated with atropine



FIGURE 2 Lateral x-ray of the head and neck showing the extent of the encephalomyelocle and reduction in number of cervical vertebrae. The hydrocephalic changes of the calvarium are evident.

$0.02 \text{ mg} \cdot \text{kg}^{-1}$  after fasting for four hours (the last feed being  $30 \text{ ml}$  of five per cent dextrose). Prophylactic antibiotic coverage started on the day of surgery and consisted of aqueous penicillin  $100,000$  units given one hour before surgery.

The baby was brought to the operating room and great care was taken while transferring him to the operating table. He was placed on his side on a heating blanket and temperature was monitored by means of a rectal thermistor probe. Blood pressure was measured every five minutes using an electronic oscillonometer (Dinamap). The ECG was monitored continuously. An oesophageal stethoscope was introduced. A vein in the foot was cannulated and inhalational induction in the lateral position was carried out using 50 per cent nitrous oxide in oxygen and increasing concentrations of halothane ( $0.5\text{--}2$  per cent). One minute before laryngoscopy  $1 \text{ mg} \cdot \text{kg}^{-1}$  lidocaine was given intravenously (IV). While an assistant was holding the head and shoulders in the correct alignment, the trachea was intubated with a Rusch flexo-metallic tube, size 14 Fr., without difficulty. Halothane was discontinued after induction and anaesthesia was maintained with nitrous oxide-oxygen (1:1) and atracurium. A total of  $4 \text{ mg}$  of atracurium was used for muscle relaxation during the procedure and ventilation was controlled using the Jackson Rees modification of Ayre's T-piece with a total flow of  $3 \text{ L} \cdot \text{min}^{-1}$ . Neuromuscular activity was assessed by observation of thumb adduction using a Bard peripheral nerve stimulator supplying supramaximal stimuli to the ulnar nerve at the wrist via surface electrodes. Train of four (TOF) stimuli, produced every 12 seconds with a current output of  $30 \text{ mA}$ , were used. Blood pressure and pulse were stable and there was no hypertensive or tachycardic response to laryngoscopy and endotracheal intubation. The patient was placed in the prone position for surgery. Meticulous attention was given to the positioning of the patient. A suitable size ring was placed under the head to ensure immobilization and supported on either side by sand-bags. Transverse bolsters were placed under the chest and pelvic girdle to ensure free movements of the diaphragm. The estimated blood loss was  $80 \text{ ml}$ , which was replaced. Anaesthesia continued without problems and lasted for 60 minutes. At the end of the surgery, nitrous oxide was withdrawn and residual neuromuscular block was reversed by neostigmine  $0.05 \text{ mg} \cdot \text{kg}^{-1}$  and



FIGURE 3 CT scan of the level of C4 showing spina bifida of the cervical spine communicating with the encephalocele.

atropine 0.02 mg·kg<sup>-1</sup>. Although all four responses to TOF were present, the endotracheal tube was left in place until the baby was fully responsive and it was removed one hour later in the ICU.

#### Discussion

Patients with Klippel-Feil syndrome pose numerous problems to the anaesthetist. They have cervical instability and are at high risk for spinal cord injury during laryngoscopy, intubation and positioning for surgery. Associated abnormalities at the atlanto-occipital junction, spinal canal stenosis or scoliosis also increase the risk for neurological damage.<sup>2</sup> Therefore, neck radiographs should be done in lateral view in all patients with Klippel-Feil syndrome. Syncopal attacks may be precipitated by sudden rotatory movements of the neck in patients with Klippel-Feil syndrome.<sup>3</sup> The anaesthetist should be particularly careful to avoid forceful neck movements when managing patients with this syndrome.

The incidence of spina bifida cystica ranges from 0.2 to 4.2 per 1,000 live births,<sup>4</sup> of these one in six may have an encephalocele,<sup>4</sup> i.e., 0.33 to seven per 10,000 live births. The incidence of encephalocele with Klippel-Feil syndrome is not known, but must be very rare.

This patient had a large occipital encephalocele which made it impossible to position the baby supine therefore the head and body had to be aligned in the semilateral position during intubation.<sup>5</sup> As problems during intubation were anticipated, muscle relaxants were not used during induction of

anaesthesia. Awake intubation is considered to be contraindicated<sup>6</sup> for patients in heart failure.

Intravenous lidocaine was given prior to laryngoscopy and intubation to reduce the hypertensive and tachycardic effects of laryngoscopy.<sup>7</sup> With the infant in the prone position, ventilation had to be controlled. The use of non-depolarizing muscle relaxants is safe but neuromuscular function should be monitored, using a peripheral nerve stimulator. The decreased margin of safety for neuromuscular transmission in neonates has been described.<sup>8</sup> Atracurium is a new nondepolarizing muscle relaxant. It has a short duration of action, almost no cardiovascular side effects when given in small doses and is easily reversed with anticholinesterase drugs.<sup>9</sup> Therefore, it offers certain advantages over the other non-depolarizing relaxants.

Any reduction in neuromuscular transmission in the postoperative period may diminish the patient's ventilatory capacity. Added to this, sleep-induced ventilatory dysfunction may occur in patients with structural central nervous system lesions.<sup>10</sup> Cleft palate, mandibular malformations and microgathia have also been described in patients with Klippel-Feil syndrome<sup>11</sup> which may contribute to anatomical upper airway obstruction.<sup>12</sup> Therefore, close observation is mandatory until the patient is fully recovered from anaesthesia.

Cardiovascular anomalies have been recognized in patients with Klippel-Feil syndrome<sup>2,13,14</sup> with an incidence 4.2 to 14 per cent.<sup>2,15,16</sup> German measles has been implicated with the development of patent ductus arteriosus<sup>17</sup> but the absence of rubella-specific IgM in this baby excluded congenital rubella syndrome. The presence of patent ductus arteriosus is of special significance to anaesthetists as it may precipitate left ventricular failure within the first few weeks of life. This neonate had heart failure treated with digitalis and diuretics. Because of the likelihood of myocardial depression and hypotension, halothane was avoided during the maintenance of anaesthesia. The persistent ductus provides a site for infective endocarditis, therefore, prophylactic antibiotics are indicated before any surgical procedure. For a full review of the anaesthesia for patients with congenital heart disease the reader is referred to the reviews by Duncan<sup>18</sup> and by Bland and Williams.<sup>19</sup>

Skeletal abnormalities are a feature of Klippel-Feil syndrome. In one series, 60 per cent of

patients exhibited significant scoliosis.<sup>20</sup> In a retrospective analysis of 21 patients with Klippel-Feil syndrome, Nagib *et al.*<sup>21</sup> reported that nine patients developed neurological deficits either spontaneously or after minor trauma. Skull asymmetry, platybasia, basilar invagination and brachycephaly have also been reported with Klippel-Feil syndrome.<sup>2,22</sup> This patient had Sprengel's deformity, spina bifida in all cervical and upper dorsal spines and fusion of C<sub>2-3</sub> vertebrae (Type II) which is thought to be autosomal-recessive<sup>22</sup> and this is the most commonly seen type of Klippel-Feil syndrome. Type I is applied to patients with extensive cervical and upper thoracic fusion.

Patients with Klippel-Feil syndrome may require general anaesthesia for correction of associated congenital anomalies or for surgical stabilization of the cervical spine. A successful outcome requires an understanding of the anatomical and pathophysiological changes associated with this syndrome. Therefore, careful preoperative examination is essential to exclude associated congenital anomalies. It can not be overstressed that manipulation of the neck during intubation and thereafter must be carefully controlled if neurological damage is to be avoided.

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#### Résumé

*L'anesthésie pour la répartition d'une encéphalomyocèle occipito cervicale chez un nouveau-né atteint d'un syndrome Klippel-Feil est décrite. La fusion de la vertèbre cervicale, un cou court, une insertion postérieure basse de la ligne du cuir chevelu et une déformation de Sprengel's, présent chez ce patient signes le syndrome de Klippel-Feil. En plus des signes habituelles du syndrome ce patient avait un large encéphalomyocèle ainsi qu'un canal artériel persistant compliqué d'une insuffisance cardiaque. Les patients atteints de ce syndrome sont susceptibles de subir des lésions de la colonne cervicale, des lésions neurologiques non seulement lors de la laryngoscopie et de l'intubation mais aussi lors de la procédure. Les implications du syndrome de Klippel-Feil pour les anesthésistes sont discutées.*