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# Anesthetic Considerations in Cri Du Chat Syndrome: A Report of Three Cases

MASAO YAMASHITA, M.D.,* FUMIO TANIOKA, M.D.,† KAZUO TANIGUCHI, M.D.,‡
AKITOMO MATSUKI, M.D.,§ TSUTOMU OYAMA, M.D.¶

Limited consideration has been given to the anesthetic management of patients with cri du chat syndrome.[1,2] This is a syndrome associated with a unique mewing quality like that of a kitten and is characterized by a partial deletion of the short-arm of chromosome number 5.[3] A recent report[4] on home-reared children indicates that the prognosis of these patients is more favorable than previously described in studies of institutionalized patients. Thus, we may see an increasing number of these patients requiring anesthesia and surgery.

We report our anesthetic experiences on three patients with cri du chat syndrome.

## REPORT OF THREE CASES

*Patient 1.* This 2,600-g infant was born to a 29-year-old mother after an uncomplicated gestation. The infant was mildly depressed at birth (Apgar 6) but responded quickly to suctioning and oxygen given via a mask. Because of characteristic facial features (microcephaly, rounded face, hypertelorism, oblique palpebral fissures, epicanthus, low-set ears, and micrognathia) and cry, the diagnosis of cri du chat syndrome was made. Subsequent chromosomal analysis confirmed this clinical diagnosis. Other findings on physical examination included the following: simian crease, unbilical hernia, prolapsus recti, dislocation of the hip, and lumbosacral meningomyelocele. The operation was scheduled for repair of the meningomyelocele at 7 weeks of age, at which time body weight was 3,890 g.

After the patient received scopolamine 0.05 mg im, anesthesia was induced and maintained with halothane, nitrous oxide, and oxygen. A 2.5-mm ID endotracheal tube was inserted into the trachea without the use of muscle relaxants. No apparent abnormalities were noticed in the larynx. Repair of lumbosacral meningomyelocele was completed without incident. The trachea was extubated uneventfully shortly after surgery. Mild respiratory distress (tachypnea, retractions) was noticed approximately 6 h later; this resolved over the next 2 h, and the remainder of the postoperative course was uneventful.

*Patient 2.* This 3,020-g product of an uncomplicated gestation was mildly asphyxiated at birth but responded to oxygen given via a mask. Shortly after birth, the infant was clinically diagnosed as having cri du chat syndrome. In addition, physical examination revealed a heart murmur. Cardiac catheterization revealed a patent ductus arteriosus (PDA) and pulmonary hypertension. The infant was scheduled for ligation of her PDA.

After premedication with scopolamine 0.03 mg im, anesthesia was induced with halothane, nitrous oxide, and oxygen. The trachea was intubated (tube size ID 3.5 mm) with the aid of succinylcholine 2.5 mg iv. Because of retromicrognathia and a long, curved, floppy epiglottis, visualization of the vocal cords was limited. For surgery the infant was placed in the right lateral position. Several minor ventilatory problems occurred during surgery: left upper lobe atelectasis, which seemed to respond to increase in peak airway pressure; and bloody secretions requiring vigorous suctioning. The trachea remained intubated for 4 h postoperatively until the cardiovascular system was stable. The postoperative chest roentgenogram was unremarkable. The infant's subsequent course was uneventful until day four, when she died after aspiration of milk, probably attributable

* Senior Lecturer, Department of Anesthesiology, University of Hirosaki School of Medicine.

† Lecturer, Department of Anesthesiology, University of Hirosaki School of Medicine.

‡ Associate Professor, Department of Anesthesiology, Ohita Medical College.

§ Associate Professor, Department of Anesthesiology, University of Hirosaki School of Medicine.

¶ Professor, Department of Anesthesiology, University of Hirosaki School of Medicine.

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Address reprint requests to Dr. Yamashita.

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to laryngeal incompetence. Autopsy revealed an atrial septal defect (ASD), a ventricular septal defect (VSD), and a ligated ductus arteriosus.

*Patient 3.* This infant's birth weight was 2,660 g, and cat-like crying, poor sucking, failure to thrive, and frequent respiratory infections characterized her infantile period. Clinical diagnosis of cri du chat syndrome was confirmed by chromosomal study. In addition to the common findings in this syndrome (abnormal cry, micrognathia, hypertelorism, mental retardation), she had a scoliosis, a mild tracheal stenosis at the seventh cervical vertebra, and a double pelvis of her right kidney. On physical examination, she had heart murmurs consistent with a VSD and pulmonary hypertension. The diagnosis was confirmed by cardiac catheterization, and she was scheduled for closure of her VSD at age four years, at which time body weight was 15.1 kg and height 115 cm.

After premedication with scopolamine 0.1 mg and morphine 2 mg im, anesthesia was induced with thiopental iv. A 5.0-mm endotracheal tube was inserted into the trachea over a modified Miller blade (Matsuki's blade[5]) after administration of iv succinylcholine. Because there was a large leak around this tube, it was replaced with a 5.5-mm tube. No apparent abnormalities of the larynx were found during intubation. Anesthesia was maintained with halothane, nitrous oxide, oxygen, and pancuronium. Although occasional premature ventricular contractions were observed at sternotomy and weaning from the cardiopulmonary bypass, anesthetic course was uneventful. The VSD was closed without complications. The postoperative course was uneventful.

## Discussion

In 1963, Lejeune *et al.*[6] reported the presence of the following features in three children: severe mental and physical retardation, microcephaly, hypertelorism, epicanthal folds, microretrognathia, low-set ears, abnormal dermatoglyphics, and, in particular, a high-pitched, plaintive cry similar to the mewing of a cat. This new clinical entity later was labeled cri du chat syndrome; it is also called "cat cry syndrome," "5P-syndrome," or "del (5P) syndrome."[3,7] The underlying chromosomal aberration was found to be partial deletion of the short arm of chromosome number 5.[7] The incidence of the syndrome is estimated to be one in 50,000, and the prevalence among mentally retarded people with an IQ below 50 is one in 350, with a male-to-female ratio of 0.72.[8] At birth, these infants are small (average birth weight 2,650 g[3]) and often are asphyxiated.[8] Cyanotic episodes, inspiratory stridor, and feeding difficulties are common. Pneumonia, aspiration pneumonia, congenital heart defects, and respiratory distress syndrome are the most frequent causes of death in the neonatal period.[8]

Physical and mental development is retarded; most children have an IQ below 50, and weight and height always remain below normal.[8]

Pathogenesis of the cat-like cry is not clear. Although abnormalities of the larynx are very common in cri du chat syndrome, they are not always present nor are the abnormalities always the same.[8] Therefore, the abnormal cry cannot be related purely to abnormalities of the larynx. Because some patients with this characteristic cry have very minor or no laryngeal anomalies,[8] the pathogenesis must be more complicated. A functional or organic neurologic defect probably contributes significantly to the pathogenesis of this cry.

The reported laryngeal abnormalities include the following: a small larynx,[6] and narrow diamond-shaped larynx,[9] laryngomalacia,[6] and vocal cord paralysis (one case reported[10]). In addition, many specific abnormalities of the epiglottis have been described: long, curved, and floppy[2]; small and flaccid[10]; hypoplastic; and hypotonic.[10] We observed a long, floppy epiglottis in our second patient.

In addition to these laryngeal abnormalities, retrognathia and a high palatal vault may make intubation difficult. Endotracheal tubes and laryngoscope blades of various sizes and shapes[5] should be available for intubation. Because many infants are hypotonic,[8] the response to muscle relaxants may be accentuated and unpredictable. This places emphasis on giving small doses and monitoring neuromuscular function.

The high incidence of congenital heart disease[8] and chronic respiratory infections from aspiration[8] introduce additional risks for anesthesia and, at times, imply intense postoperative care. We anesthetized our three patients successfully with halothane and nitrous oxide in oxygen. However, there is no particular contraindication to choosing either other inhaled or iv anesthetics.

In summary, the main concern for anesthesiologists caring for children with this syndrome are abnormalities of the larynx and upper airway, congenital heart disease, and mental retardation. With proper attention to these factors, however, successful anesthetic management with conventional anesthetic techniques can be accomplished.

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