Anaesthetic management of Miller's syndrome

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Miller's syndrome is a rare congenital disorder with facial features similar to that of Treacher-Collins syndrome. This report details the anaesthetic management of an infant during multiple surgical procedures, beginning with pyloromyotomy at one month of age. Airway management was difficult because of severe micrognathia and was accomplished using an awake intubation with a conventional straight blade modified for continuous administration of oxygen ("oxyscope"). Due to recurrent upper airway obstruction and the anticipated need for multiple surgical procedures in the first years of life, a tracheostomy was placed. Because of the multiple airway, orthopaedic, and nutritional difficulties, it is important that a prospective, multidisciplinary approach be used in these patients' care. Consideration should be given to early tracheostomy for airway maintenance.

Le syndrome de Miller est une maladie congénitale rare avec des caractéristiques faciales similaires au syndrome de Treacher-Collins. Ce rapport étudie la conduite anesthésique chez un enfant lors de procédures chirurgicales multiples, commençant par une pyloromyotomie à l'âge d'un mois. Le contrôle des voies aériennes fut difficile à cause d'une micrognathie et fut accompli par une intubation réveillée avec une lame droite conventionnelle modifiée pour l'administration continue d'oxygène (« oxyscope »). À cause de problèmes d'obstruction des voies aériennes hautes récurrentes et l'anticipation d'une nécessité de procédures chirurgicales multiples dans les premières années de la vie, une trachéotomie fut faite. À cause de problèmes multiples des voies aériennes, orthopédique et nutritionnelles, il est important d'instaurer une approche

multidisciplinaire pour le soin de ces patients. Des considérations doivent être faites pour une trachéotomie précoce pour le maintien des voies aériennes.

A syndrome of postaxial acrofacial dysostosis was first described by Miller in 1979.¹ This rare congenital syndrome consists of multiple facial abnormalities (micrognathia, malar hypoplasia, eyelid coloboma, low-set and cupped ears, cleft lip and/or palate) and severe deformities of the limbs.¹⁻⁷ To promote psychosocial adjustment, surgical correction of cosmetic defects is indicated. Multiple surgical procedures spanning infancy, adolescence, and adulthood can be anticipated. It is important that both paediatric and adult anaesthetists be familiar with the anesthetic implications of this syndrome.

Case report

This one-month-old 2.9 kg child was diagnosed shortly after birth as Miller's syndrome. Poor weight gain and intermittent partial upper airway obstruction resulted from severe micrognathia that required continuous observation in the neonatal intensive care unit (NICU). Projectile vomiting began at four weeks of age. Radiological and ultrasound examination suggested pyloric stenosis. Preoperative physical examination revealed characteristic craniofacial abnormalities and limb defects of Miller's syndrome (Figure). Micrognathia was pronounced; an oral airway was taped in place. Chest and heart examinations, urinalysis, electrolytes, BUN, and creatinine concentrations were normal. Echocardiogram was also normal. Atropine 0.09 mg, was given *im* 45 min before surgery. With intravenous access in place, we attempted an awake intubation with a #0 Miller oxyscope (a straight blade modified with an oxygen channel extending to the tip); the larynx could not be visualized. After several attempts with a #1 Miller oxyscope, the extreme posterior aspect of the larynx was visualized and a 3.0 mm oral endotracheal tube was placed. Proper positioning of the tube was confirmed by chest auscultation and capnography. Anaesthesia was induced with halothane in inspired concentrations up to 1.5%, but the systolic blood pressure decreased from 80 to 50 mmHg. Anaesthesia was subsequently maintained with 0.2%

Key words

ANAESTHESIA: paediatric;
SYNDROMES: Miller's.

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FIGURE The patient aged 11 months. Note retruded brow, bilateral lid colobomata, low-set cupped ears, micrognathia, and shortened limb with postaxial deformities. Also note tracheostomy and feeding tubes are in place.

halothane in oxygen, in addition to 0.5% bupivacaine injected at the surgical site. Neuromuscular blockade was achieved with vecuronium. At the conclusion of the pyloromyotomy, neuromuscular blockade was antagonized, and the trachea was extubated when the child was awake. After extubation, the child required replacement of the oral airway to relieve partial upper airway obstruction. The child remained in the NICU both for observation of respiratory status and nutritional support. During the next few days, the child was further evaluated by plastic surgery, ophthalmology, audiology, orthopaedic, and speech pathology services. Repeat echocardiogram revealed a small patent ductus arteriosus which did not require treatment. The otolaryngology and neonatology services monitored the patient's respiratory status. There was a slowly increasing degree of upper airway obstruction associated with increasing dependence on the presence of an oral airway. The anticipated need for multiple surgical procedures during the first few months of life was

discussed and tracheostomy was performed. The 3 kg child received premedication 20 min before surgery with 0.1 mg of atropine *im*. With intravenous access and monitoring in place, awake laryngoscopy with a Holinger laryngoscope (a straight blade laryngoscopy modified with an open slot on the right side of the instrument; Pilling Endoscopic Instruments, Fort Washington PA 19034) was performed. Visualization was again difficult, but full exposure was obtained using a right lateral approach to larynx (by sliding the laryngoscope blade down the extreme right side of mouth, keeping the tongue to the far left). The otolaryngologist passed a 2.5 mm $\times$ 20 cm ventilating bronchoscope which was left in place. Anaesthesia was maintained with 0.5% isoflurane and oxygen and relaxation was produced with vecuronium. After completion of the 90 min procedure (including bilateral myringotomy and tube placement), neuromuscular reversal was accomplished, and the child returned to the NICU breathing spontaneously through a 00 Shiley neonatal tracheostomy tube. The child remained in hospital for one month after the tracheostomy both for nutrition support and conservative treatment of hip dislocation. She returned to the operating room several times during the next six months for procedures including reduction of hip dislocation, hip spica cast changes, correction of eyelid colobomas, bronchoscopy, cleft palate closure, Nissen fundoplication, and gastrostomy. Anaesthetic management and patient safety was facilitated by the presence of the tracheostomy tube. Anaesthetics for these procedures consisted of volatile anaesthetics combined with neuromuscular relaxants. The only major anaesthetic-related problem was difficulty of venous access which was accomplished with a central (external jugular) approach. Major reconstructive extremity procedures, as well as craniofacial reconstruction, are planned in the future. Tracheostomy removal is anticipated after a mandibular lengthening procedure.

Discussion

The syndrome currently known as Miller's syndrome or acrofacial dysostosis was first defined by Miller in 1979, but had previously been referred to as the Genee-Wiedemann syndrome.^{1,3} It is extremely rare, with fewer than 20 cases reported. Miller's syndrome is characterized by facial features that are very similar to the more common Treacher-Collins syndrome, but with the addition of limb abnormalities not seen in that syndrome.^{5,8} It is also similar to Nager's syndrome, except that the latter has preaxial extremity defects instead of postaxial.⁵ Miller's syndrome is primarily characterized by its craniofacial and limb abnormalities; these patients have a high frequency of various associated defects including an approximately 30% incidence of cardiac disease (Table).

TABLE Miller's syndrome—recognized features¹⁻⁷

<i>Craniofacial</i>
Malar hypoplasia*
Lateral orbital cleft*
Superolateral retruded brow*
Coloboma lower lid*
Micrognathia*
Cleft lip and/or palate
Cupped ears
<i>Cardiothoracic</i>
Congenital heart disease (atrial and ventricular septal defects, patent ductus arteriosus)
<i>Limbs</i>
Postaxial deformities (i.e., ulnar aspect)*
Absence of 4th and 5th rays in hands and feet*
Shortening of limbs*
<i>Neurological</i>
Usually normal intelligence
Hearing deficit
<i>Miscellaneous</i>
Accessory nipples
Cryptorchidism
Difficult venous access
Gastro-oesophageal reflux

*Universal features.

The degree of penetrance of this syndrome is quite variable in the extent of both facial and limb defects.

The pathophysiology of Miller's syndrome is not yet firmly established. The syndrome has been reported in siblings; an autosomal recessive mode of inheritance has been postulated.^{2,4,6}

Mental development in these patients is generally normal.^{1,4} Multiple plastic surgical and orthopaedic procedures are appropriate to encourage normal psychosocial development. The anaesthetic management required for these procedures is poorly described; with one exception, all previous case reports appear in the non-anaesthesia literature. The anaesthetic management was described by Richards in 1987.⁹ He anaesthetised a 12-month-old child with this syndrome for repair of a cleft palate. The primary problem during anaesthetic management was difficult tracheal intubation due to severe micrognathia. The author reported a gaseous induction (using halothane and oxygen) which resulted in complete airway obstruction. After giving succinylcholine, blind placement of an introducer into the trachea was accomplished, over which an endotracheal tube was subsequently guided. After the surgical procedure, the trachea was extubated while the child was "deeply anaesthetized." Postoperatively, there were multiple episodes of partial airway obstruction.

The patient described in our report manifested Miller's syndrome in a severe form. Micrognathia made airway management a constant problem since birth. Airway management of this patient differed from that presented by Richards. We chose to secure the airway by awake intubation before introduction of anaesthetic agents during preparation for the pyloromyotomy and the later tracheostomy. Although laryngeal visualization was difficult in both anaesthetics, control of the airway was obtained by direct visualization of the larynx. Had direct laryngoscopy not been successful, alternatives for intubation were at hand (including a fiberoptic laryngoscope, equipment for cricothyroidotomy, and means for trans-tracheal jet ventilation). To minimize the chances of airway obstruction the trachea was extubated at the end of surgical procedure only when the patient was fully awake.

We made an aggressive effort both before and after the patient's original surgical procedure to maintain the airway without the need for tracheostomy. The decision to proceed with a tracheostomy followed discussion between the surgical and medical subspecialties involved in this child's treatment, with consideration for the multiple surgical procedures planned in early life. No doubt the safety of anaesthesia management during subsequent surgical procedures was facilitated by the presence of tracheostomy.

The management of patients with Miller's syndrome involves members of many subspecialties including anaesthesia. Patient outcome can be maximized through communication between the physicians and the family of the patient. Anaesthetic care of a patient with Miller's syndrome is complicated by venous access, potential for cardiac defects, and airway management. Airway management is similar to that of the more common Teacher-Collins syndrome, but because patients with Miller's syndrome are likely to require multiple surgical interventions during infancy, early tracheostomy should be considered.

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