Anaesthetic management of children with Moebius syndrome

T.M. Shichor, M. Speakman, W.A. Ames, R.M. Zuker, C. McCaul
Departments of Anaesthesia and Plastic Surgery, The Hospital for Sick Children, Toronto, Ontario

Introduction: Moebius syndrome is a rare, complex congenital anomaly characterized by facial nerve palsy, other cranial neuropathies, limb abnormalities, orofacial malformations and developmental delay. Children with Moebius syndrome may present for surgical correction of strabismus, cleft palate, limb malformations as well as conditions unrelated to the syndrome itself. We present our experience of anaesthesia for children with Moebius syndrome.

Methods: With institutional ethics board approval, the charts of all patients presenting for gracilis muscle transplant surgery between January 1994 and January 2003 were reviewed retrospectively. All patients with a clinical diagnosis of Moebius syndrome (N=47) were included in the study, and all anaesthetics they had undergone at our institution were reviewed.

Results:

Demographics and clinical features: The male-to-female ratio was 1:1.75 and age at the time of surgery ranged from 1 month to 18.5 years (mean 8 years). Facial nerve palsy was present in all patients and bilateral in 44 (93.6%). 32 patients (68%) had concomitant 6th cranial nerve palsy and 20 (42%) had involvement of other cranial nerves. Limb defects (clubfoot, syndactyly and congenital amputations) were present in 21 patients (44.6%), orofacial anomalies (retrognathia, mandibular hypoplasia, cleft palate) in 19 (40%) and developmental delay in 9 (19.1%). 4 patients had congenital cardiac defects (PDA, VSD, PDA+ASD+VSD, PDA+dextrocardia).

Anaesthesia: 111 procedures under anaesthesia were performed – 76 gracilis muscle transplants, 10 orthopedic, 9 dental, 6 ENT/ophthalmology and 10 others. The mean duration of anaesthesia was 6.8 hours (range 0.3-12.25). Inhalation induction was used in 65 cases (58.5%) and intravenous in 46 (41.5%). For maintenance, volatile anaesthetics (halothane/isoflurane) were used in all patients. Muscle relaxants were administered in 87 cases (78.4%) and opioids in 90 (81.8%).

Airway management: Mask ventilation was easy in all patients. Endotracheal intubation was performed in 106 cases and was difficult in 35 cases (28.3%) in 6 patients (12.8%). In these cases, intubation was aided by cricoid pressure alone in 17 cases (16%). In 13 cases (12.6%) a combination of measures was needed to facilitate intubation, including cricoid pressure, stylettes, two-person technique, and changing of the laryngoscope blade. Fiberoptic intubation was performed in 3 cases, a reinforced laryngeal mask used in one, and one procedure was cancelled due to failed intubation. All intubated patients were extubated at the end of surgery.

Analagesia: Intra- and postoperative pain management varied widely and included morphine by bolus, infusion or PCA, fentanyl or meperidine PCA, codeine and paracetamol. Epidural analgesia was used in one patient. There were no complications related to pain management.

Complications: All intraoperative courses were uneventful. One patient was re-intubated due to apnea and extubated successfully in the PACU, and there were two cases of post-extubation stridor. Oxygen therapy was required beyond the immediate postoperative period in 9 cases. There were two cases of pneumonia, one atelectasis, and two cases of wheezing and stridor. No patient required ventilatory support and there were no ICU admissions. Of 14 ambulatory procedures there were no unplanned hospital admissions.

Discussion: Airway difficulties were the most frequently encountered anaesthetic challenge. The incidence of difficult intubation in our series, while lower than previously reported, warrants a high degree of clinical suspicion. Simple maneuvers such as cricoid pressure can be used to facilitate successful intubation in the majority of patients. Intubation failure is rare. There was no significant morbidity associated with prolonged anaesthesia, and with the use of opioids or muscle relaxants. We conclude that anaesthesia for patients with Moebius syndrome can be performed safely and with minimal complications, including ambulatory surgery in selected cases.

Refs.

1. Stromland K. et al., Eur J of Pediatr Neurol 2002
2. Ferguson S., Paediatr Anaesth 1996