

## Anaesthesia recommendations for patients suffering from **Moebius syndrome**

**Disease name:** Moebius syndrome

**ICD 10:** Q87.0

**OMIM:** 157900

**Synonyms:** Congenital facial diplegia (Congenital oculofacial paralysis), Möbius syndrome, Moebius sequence, MBS

Moebius syndrome is a rare, nonprogressive neurological disorder (prevalence is estimated to be 0.002% of births) characterized by unilateral or bilateral facial paralysis and defective extraocular eye movements secondary to congenital paresis of the facial (VII) and abducens (VI) cranial nerves. These classic features of the syndrome are often accompanied by hypoglossal (XII), trigeminal (V), glossopharyngeal (IX) and vagal (X) nerve palsies. Affected infants typically present with congenital esotropia and immobile, expressionless facies. Depending on the pattern of cranial nerve involvement, there may be a wide range of clinical expression. Feeding difficulties due to poor coordination of sucking and swallowing may be present with IXth and Xth cranial nerve involvement. This may be associated with dysphagia and retention of oral secretions leading to recurrent bouts of aspiration pneumonia. Inadequate function of the soft palate can also result in dysarthria. Moebius syndrome may also occur in association with various craniofacial (mandibular hypoplasia, microstomia, temporomandibular joint dysfunction, cleft palate, external ear deformities), limb (club foot) and musculoskeletal malformations as well as multiple ophthalmic abnormalities (incomplete eye closure, inability to blink). Other associated manifestations include seizure disorders, congenital heart diseases, hypotonia, hypogonadotropic hypogonadism, hydrosyringomyelia and some degree of mental retardation. It is also associated with prematurity.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)

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## Disease summary

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The cause of Moebius syndrome is unknown, but rhombencephalic maldevelopment and brainstem ischemia during the first trimester are two possible etiological hypotheses in children with normal karyotype. The list of potential associated teratogenic events has included hyperthermia, trauma, thrombus formation, embolism, hemorrhage, as well as in utero exposure to various medications including misoprostol. Most cases are sporadic, but some familial cases are also known. The inheritance patterns of Moebius sequence are heterogeneous and can be autosomal recessive, autosomal dominant or even X-linked. Some candidate regions and candidate genes (3q21-q22 and 13q12.2-q13) have been described but no causative gene has yet been confirmed.

The syndrome has most frequently been confused with hereditary congenital facial palsy which is restricted to involvement of the facial nerve and no other abnormalities.

Poland-Mobius syndrome is a rare congenital disorder that includes combination features of Poland and Mobius syndromes. Poland syndrome consists of absence of pectoralis major muscle, syndactyly, brachydactyly and hypoplasia of the hands.

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## Typical surgery

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Anesthesia for imaging (CAT scan and MRI) and surgical procedures that may include correction of eye anomalies (e.g. strabismus surgery, ptosis repair, tarsorrhaphy), orthopedic problems (correction of limb abnormalities), plastic/reconstructive (cleft palate, jaw surgery, facial reanimation surgery), otolaryngological, dental (teeth extractions), or general surgical interventions.

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## Type of anaesthesia

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The potential for problems with aspiration of oral secretions should be remembered and the use of antisialogogue premedication is recommended.

General anaesthesia is potentially high-risk, due to the airway management difficulties.

Regional anesthesia per se is not contraindicated, however, positioning problems and scoliosis can affect the success rate negatively.

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## Necessary additional diagnostic procedures (preoperative)

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Although a literature review did not identify reports of an association of cardiomyopathy with Moebius syndrome there are suggestions of myocardial involvement (ventricular septal defect, ductus arteriosus, dextrocardia) thereby emphasizing the consideration of preoperative echocardiography in these patients if one has not been previously obtained.

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### **Particular preparation for airway management**

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Craniofacial changes, which can hinder tracheal intubation considerably, are seen in approximately 90% of the patients. Micrognathia, retrognathia, mandibular hypoplasia, and cleft palate are some of the manifestations seen in these patients.

Anaesthesia using a facemask with or without a Guedel airway appeared to be well tolerated and is probably the method of choice where appropriate. Face mask ventilation is usually adequate.

Combination of measures could be used to facilitate intubation, including cricoid pressure, stylettes, gum elastic bougie, two-person technique, changing of the laryngoscope blade and fiberoptic scope.

In a case of failed intubation the airway could be managed with a laryngeal mask, although its placement could also be difficult (has been reported to have a high failure rate).

Difficult airway equipment should be available.

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### **Particular preparation for transfusion or administration of blood products**

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None known or reported.

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### **Particular preparation for anticoagulation**

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None known or reported.

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### **Particular precautions for positioning, transport or mobilisation**

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Care should be taken to position and protect all affected limbs in the neutral position to reduce the risk of pressure areas or neuropraxia.

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### **Probable interaction between anaesthetic agents and patient's long term medication**

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Although seizures are not a consistent finding in patients with Moebius syndrome, there are reports of associated epilepsy. Anesthetic care in patients with a seizure disorder should include the recent documentation of serum anticonvulsant concentrations to ensure therapeutic dosing. Continuation of anticonvulsant medications to maintain therapeutic levels intraoperatively is of significant concern as is the immediate reinstatement of chronic seizure therapy during the postoperative period. The induction of hepatic enzymes by certain anticonvulsants may alter the pharmacokinetics and pharmacodynamics of several drugs, including neuromuscular blocking agents (NMBAs). Increasing the intraoperative doses of NMBAs and certain intravenous anesthetic induction agents may be necessary with concomitant anticonvulsant therapy.

## **Anaesthesiologic procedure**

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Associated mental retardation and visual and hearing disturbances, both of which may present a major challenge in the communication with and assessment of the patient.

Involvement of the hypoglossal nerve can lead to hypoglossia or ankyloglossia with abnormalities of tongue coordination. These may further increase the likelihood of problems with secretions. The use of antisialogogue premedication is recommended.

Facial paralysis may result in incomplete eye closure and the inability to blink, thereby placing the patient at risk for exposure keratopathy and corneal ulcerations even without anesthetic care. In such cases, meticulous attention to eye care is suggested.

Induction of anesthesia can be intravenous or inhalational.

The use of succinylcholine should be avoided because of the potential risk of rhabdomyolysis, hyperkalemia, and malignant hyperthermia. The absolute risk of malignant hyperpyrexia (MH) is unknown. A single case report of fatal MH in an infant with Moebius syndrome exists in the literature to date.

Given the association with abnormal ventilatory control, caution with opioids may be appropriate. Extended respiratory monitoring may be required.

## **Particular or additional monitoring**

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Train-of-four monitoring due to hypotonia. Electrodes should be placed at a site which is not affected by the disease process.

## **Possible complications**

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Abnormalities of the orofacial structures are common and as a result may lead to difficulties with intubation.

Potentially increased risk of regurgitation and aspiration of oral secretions or gastric contents in the perioperative period.

Secretions can cause partial airway obstruction and hypoxaemia. Respiratory failure secondary to excessive airway secretions could require postoperative mechanical ventilation.

Acute and chronic pulmonary complications which may result from aspiration.

Abnormalities of ventilatory control (apnoea, hypopnoea) presumably due to associated lesions in the brainstem.

Deformities of upper or lower limbs can be associated with difficulty in securing intravenous access.

Care should be taken to prevent corneal abrasions.

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### **Postoperative care**

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Paresis of the facial nerve and the consequent absence of facial expression impair the patient's ability to communicate nonverbally, rendering it difficult to evaluate these patients and assess their pain. Changes in physiological parameters (heart rate and blood pressure) should be used to assess level of analgesia. It is also helpful to enlist the mother's help with an assessment of child's level of comfort.

Hypotonia can also impact on postoperative respiratory function at both the level of the upper airway and the thoracic musculature and diaphragm. This is especially relevant during the perioperative period when residual anesthetic agents and NMBAs may exacerbate poor baseline function leading to respiratory failure.

Continuous postoperative monitoring of respiratory function is recommended.

For postoperative analgesia drugs with limited effects on central respiratory function should be used. Non opioid analgesics should be used preferentially.

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### **Information about emergency-like situations / Differential diagnostics**

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*caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease*

Not known or reported.

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### **Ambulatory anaesthesia**

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The ambulatory surgery should take place at a tertiary care institution with adequate resources and in selected cases.

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### **Obstetrical anaesthesia**

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Not reported.

## Literature and internet links

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*This guideline has been prepared by:*

**Authors**

**Ivana Budic**, Assist. Prof. of Anesthesiology, Centre for Anesthesia and Resuscitation, Clinical Centre Nis, Medical School University of Nis, Serbia  
[ibudic@open.telekom.rs](mailto:ibudic@open.telekom.rs)

**Marija Stevic**, Anesthesiologist, University Children's Hospital, Belgrade, Serbia  
[mamarija74@gmail.com](mailto:mamarija74@gmail.com)

**Vesna Marjanovic**, Anesthesiologist, Centre for Anesthesia and Resuscitation, Clinical Centre Nis, Serbia  
[prof.marjanovic@gmail.com](mailto:prof.marjanovic@gmail.com)

**Dusica Simic**, Prof. of Anesthesiology, University Children's Hospital, Medical School University of Belgrade, Serbia  
[dusicasimic2@gmail.com](mailto:dusicasimic2@gmail.com)

**Peer revision 1**

**Conan McCaul**, Anesthesiologist, The Rotunda Hospital Dublin, Ireland  
[cmccaul@ROTUNDA.ie](mailto:cmccaul@ROTUNDA.ie)

**Peer revision 2**

**Richard Redett**, Director Pediatric Plastic Surgery, Department of Plastic and Reconstructive Surgery, Johns Hopkins Hospital  
[rredett@jhmi.edu](mailto:rredett@jhmi.edu)

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