

Anaesthesia recommendations for **3MC syndrome / Michels syndrome**

Disease name: 3MC syndrome, Michels syndrome

ICD 10: -

Synonyms: Malpuech-Michels-Mingarelli-Carnevale syndrome

Disease summary: 3MC Syndrome, is a rare multiple anomaly disorder characterized by hypertelorism, craniosynostosis, blepharophimosis, ptosis, cleft lip/palate, genitourinary tract anomalies, umbilicadefect and caudal appendage. Mild intellectual disability and hearing loss and anterior segment dysgenesis may also be present.

3MC syndrome is the overarching name given to a group of four disorders which were originally delineated clinically as separate but overlapping entities subsequently identified to have the same genetic basis. The abbreviation 3MC therefore encompasses Mingarelli, Malpuech, Michels and Carnevale syndromes. Pathogenic variants in COLEC11, COLLEC10 and MASP1 genes may all be found in individuals with 3MC syndrome. Abnormal expression of these genes leads to impairment in the lectin complementary pathway. This pathway affects neuronal migration in many tissues, particularly in the craniofacial structures.

Medicine is in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong



Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

Cleft lip and palate repair, craniostyostosis surgery, blepharophimosis/ptosis surgery, cochlear implant, inguinal hernia repair, orchidopexy.

Type of anaesthesia

General anaesthesia.

Necessary additional pre-operative testing (beside standard care)

Spina bifida occulta must be excluded if neuroaxial blocks will be applied. Renal functions must be evaluated.

Particular preparation for airway management

Due to cranial bone deformities and cleft lip or palate, airway management may be difficult. There is only one case report in the literature in which laryngeal mask airway was applied without any difficulty. If tracheal intubation is necessary, fiberoptic intubation may be recommended, requiring an anaesthetist experienced in paediatric difficult airway management.

Particular preparation for transfusion or administration of blood products

Standard management.

Particular preparation for anticoagulation

Standard management.

Particular precautions for positioning, transportation and mobilisation

Standard management.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic procedure

There is only one case report in the literature in which laryngeal mask airway was applied under general anaesthesia. No complications were reported.

Particular or additional monitoring

Standard management.

Possible complications

Airway management problems.

Post-operative care

Not reported.

Disease-related acute problems and effect on anaesthesia and recovery

None.

Ambulatory anaesthesia

Not reported. Due to the syndromes aspects, ambulatory anaesthesia for outpatient surgery does not seem appropriate.

Obstetrical anaesthesia

Not reported.

References

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