

orphananesthesia

Anaesthesia recommendations for patients suffering from **Hallermann-Streiff syndrome**

Disease name: Hallermann-Streiff syndrome

ICD 10: Q87.8

Synonyms: Francois dyscephaly syndrome, oculo mandibulo dyscephaly

Cardinal features of this syndrome are dyscephaly with bird facies, frontal/parietal bossing, dehiscence of sutures with open fontanellae, hypotrichosis of scalp, eyebrows and eyelashes, cutaneous atrophy of scalp and nose, microphthalmia, congenital cataracts, blue sclera, nystagmus, mandibular hypoplasia, forward displacement of temporomandibular joints, high-arched palate, small mouth, multiple dental anomalies and proportionate small stature.

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Typical surgery

Ophthalmic surgeries (for congenital cataract). This will be in the neonatal period.

Type of anaesthesia

General anaesthesia.

Necessary additional diagnostic procedures (preoperative)

Usually needs a consultation by a geneticist or a paediatrician to arrive at the diagnosis.

Particular preparation for airway management

Preparedness for securing the airway while preserving spontaneous respiration.

Particular preparation for transfusion or administration of blood products

None.

Particular preparation for anticoagulation

None.

Particular precautions for positioning, transport or mobilisation

None.

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

None.

Anaesthesiologic procedure

General endotracheal anaesthesia following inhalational induction of anaesthesia. Use of Airtraq facilitates better glottic view for intubation.

Particular or additional monitoring

None.

Possible complications

Failed intubation, possibility of laryngospasm if airway is instrumented in lighter planes of anaesthesia.

Postoperative care

As in any other case.

Information about emergency-like situations / Differential diagnostics

None.

Ambulatory anaesthesia

Safe.

Obstetrical anaesthesia

Not reported / no personal experience.

Last date of modification: October 2013

This guideline has been prepared by:

Author

Handattu Krishna, Anaesthesiologist, Kasturba Medical College Manipal, India
hmkrishna20032002@gmail.com

Peer revision 1

Ian James, Anaesthesiologist, Great Ormond Street Hospital for Children, London, United Kingdom
Ian.James@gosh.nhs.uk

Peer revision 2

Jayakar Thomas, Department of Dermatology, Sree Balaji Medical College, Chromepet, Chennai, India
jayakarthomas@gmail.com
