

Anaesthesia recommendations for **Hallermann-Streiff syndrome**

Disease name: Hallermann-Streiff syndrome (HSS)

ICD 10: Q87.8

Synonyms: Francois dyscephaly syndrome, oculo-mandibular dyscephaly

Disease summary: Hallermann-Streiff syndrome is a very rare disorder. Cardinal features include craniofacial dysmorphism and upper airway abnormalities.

A bird-like facies, cutaneous atrophy of scalp and nose, hypoplastic nasal foramina, frontal/parietal bossing, dehiscence of sutures with open fontanellae, hypotrichosis of scalp, eyebrows and eyelashes, microphthalmia, congenital cataracts, blue sclera, nystagmus, mandibular hypoplasia, forward displacement of temporomandibular joints, high-arched palate, small mouth opening and multiple dental anomalies are archetypical for this syndrome.

Patients present with proportionate small stature, scoliosis and hyperextensible joints. Obstructive Sleep Apnoea, tracheomalacia, recurring bronchopulmonary infections and cor pulmonale may be present. Can be associated with congenital heart defects. Mental development may be impaired in some cases. Can also be associated with ADHS and epilepsy.

No causative treatment available.

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

Corrective ophthalmic, ENT, dental, maxillo-facial or orthopaedic surgeries. Usually in infancy.

Type of anaesthesia

Both general and regional anaesthesia are feasible. Dysmorphic features present challenges to both techniques.

Necessary additional pre-operative testing (beside standard care)

Investigation, including imaging and ENT consult, of likely difficult airway.

Look for current bronchopulmonary infection.

If congenital heart defect is present (or suspected): Echocardiography and ECG advised.

Particular preparation for airway management

A variety of reasons for a difficult airway is present in HSS. With increasing age, the difficulty in airway management seems to increase. Diligence in the preparation of airway management is advised. Successful use of a video laryngoscope under spontaneous breathing induction has been described.

However, there are reports of difficulties with any modality of airway management including mask-bag-ventilation, direct and indirect (video-enhanced) laryngoscopy as well as nasal intubation.

Consider primary tracheostomy in the most severe cases.

Particular preparation for transfusion or administration of blood products

No data available.

Particular preparation for anticoagulation

No data available.

Particular precautions for positioning, transportation and mobilisation

Hyperextensible joints, slender features and possible scoliosis demand special attention to positioning.

Interactions of chronic disease and anaesthesia medications

Interactions with anti-epileptics are to be considered. Peri-operative continuation of these drugs is recommended.

Anaesthetic procedure

Regional anaesthesia may be a good alternative in some surgeries as it should be possible to preserve spontaneous breathing.

Should general anaesthesia be necessary, securing the airway will be the most difficult aspect of the procedure.

Particular or additional monitoring

Extended cardiovascular monitoring in cases of congenital heart disease.

Possible complications

“Cannot intubate-cannot ventilate” situations must be avoided through proper airway management.

Post-operative care

Airway obstruction and sleep apnoea may complicate post-operative course. Prolonged saturation monitoring is advisable. Surgeries involving the airway must be considered for their potential effects on ventilation.

Disease-related acute problems and effect on anaesthesia and recovery

Not reported.

Ambulatory anaesthesia

Possible. Surgery must not involve airway. Adequate duration of monitoring in the recovery period must be provided.

Obstetrical anaesthesia

Not reported.

References

1. Malde AD, Jagtap SR, Pantvaidya SH. Hallermann-Streiff syndrome: airway problems during anaesthesia. *J Postgrad Med* 1994 Oct-Dec;40(4):216–218
2. Ravindran R, Stoops CM. Anesthetic management of a patient with Hallermann-Streiff syndrome. *Anesth Analg* 1979;58(3):254–255
3. A Bösenberg, R Brown. Anaesthesia and Hallermann-Streiff Syndrome. *Southern African Journal of Anaesthesia & Analgesia* 2006;12(2): S5–S6
4. Krishna HM, Bhagat S, Vinodhadevi V. Difficult intubation in an infant with Hallermann–Streiff syndrome – easy with Airtraq laryngoscope. *Pediatric Anesthesia* 2012; 22:497–498. DOI:10.1111/j.1460-9592.2011.03762.x
5. Srinivasan LP, Viswanathan J. Hallermann-Streiff Syndrome: Difficulty in airway increases with increasing age. Sankara Nethralaya Medical Research Foundation, India. *Journal of Clinical Anesthesia*, 2018-11-01,50:1.

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