

Anaesthesia and Hallermann-Streiff Syndrome

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Synopsis of the patient

A 3-month old, 2.5kg female ex-premature infant presented with feeding difficulties, failure to thrive and recurrent chest infections, secondary to reflux and aspiration. She was scheduled for a Nissen fundoplication. Her dysmorphic facial features included a broad forehead, microstomia, micrognathia, low set ears and microphthalmia (Fig 1). She had no teeth. Heart sounds were normal and there were scattered rhonchi on auscultation of her chest. Chest X-ray revealed evidence of resolving aspiration pneumonia. Pregnancy was uneventful. There was no significant family history or consanguinity.

Anaesthesia was uneventful. Airway patency was easily maintained during a sevoflurane induction using a softseal facemask and continuous positive airway pressure ventilation (CPAP) despite the initial concerns that the airway would be challenging. The epiglottis was just visible at laryngoscopy with a straight Miller blade size 0, and she was successfully intubated using a stylet with cricoid pressure. Intra- and postoperative analgesia was provided by a continuous epidural infusion of 0.2% bupivacaine via a catheter placed in the T11-12 interspace. The immediate postoperative course was uneventful but the fundoplication was complicated by poor healing.

A redo Nissen fundoplication was performed at 7 months and a feeding gastrostomy was inserted at the same time. Again the immediate postoperative course was uneventful but poor healing led to problems with prolapse and herniation of the gastrostomy. She has been readmitted with recurrent chest infections. She is now 20 months old and weighs 9.2 kg.

Introduction

Hallermann-Streiff syndrome is a rare oculomandibulofacial dyscephaly (malformation of cranial and facial bones) with hypotrichosis (sparse hair), that occurs as a sporadic mutation. Although the syndrome was previously considered to have an autosomal dominant inheritance, all cases to date have been sporadic mutations.¹ The syndrome was first described by Audry in 1893.² Approximately 150 cases are recorded in the

medical literature with only 5 in anaesthesia journals.³⁻⁶ Hallermann and Streiff later independently described the clinical features in 1948 and 1950 respectively⁷, hence the name.

Clinical Features

The syndrome is characterised by malformation of cranial and facial bones, birdlike facies, sparse hair and skin atrophy, which is confined to the head and neck, microphthalmia, congenital cataracts, a beaked nose, micrognathia, dental

Figure 1: Some of the typical facial features are not obvious in infancy. However the microphthalmia and microstomia, (with thin lips and everted lower lip) and low set ears are clearly seen in this infant. The sagging "double-chin" is not obvious. A naso-jejunal feeding tube was used in an attempt to reduce the aspiration risk prior to the fundoplication.



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anomalies and proportionate short stature.^{1,7} Many of these are not obvious in the newborn but become more obvious with age and growth.

The usual facial features are frontal-parietal bossing as a result of delayed ossification of the sutures in a thin calvarium. A variety of other dycephalies have been described. The skin over the nose and skull sutures is atrophic and the hair thin with hypotrichosis on the scalp. The disproportionately small nose is thin, small and pointed because of hypoplasia of the cartilage. The nose becomes beak-shaped and parrot-like with age but is not obvious in infancy. Natal teeth may be present. These children may present for dental or orthodontic surgery for supernumerary teeth, malimplantation, hypodontia, malocclusion and dental caries.

The ocular abnormalities are the most striking. 80% have bilateral microphthalmia and 94% have cataracts that may be total or incomplete. Some may resorb spontaneously but the rest often require surgery. Despite surgery, many are destined to be blind as a result of these ocular defects. Blue sclerae have been described. Other rarer eye problems include optic disk coloboma, glaucoma, and various chorioretinal pigment changes. The eyebrows and eyelashes are usually sparse.

These children usually have normal intelligence, although mental retardation may be present in up to 15%. Epilepsy has also been described.

Airway management

Securing the airway in these children may be difficult for a number of reasons. The microstomia and mandibular hypoplasia together with the dental anomalies make laryngoscopy difficult. A high-arched palate, a particularly anteriorly situated larynx and a narrow oropharynx may compound the problem. The hypoplastic nose and a deviated nasal septum may preclude nasal intubation. The abnormal hypoplastic temporomandibular (TM) joints, if present, may dislocate during laryngoscopy unless appropriate care is taken.

The narrow upper airway, as a result of the craniofacial abnormality combined with tracheomalacia, may lead to significant respiratory complications. These include feeding difficulties and an increased risk of aspiration, chronic upper

airway obstruction, respiratory insufficiency and corpulmonale. Elective tracheostomy has been reported in some infants.

Cardiac anomalies may be present in 4.8% of these children and range from a patent ductus arteriosus, septal defects or more complex problems such as tetralogy of Fallot. In these cases prophylactic antibiotics are indicated.

Radiological features include poor ossification of the skull with wormian bones and large fontanelles, rib hypoplasia and thin long bones, with metaphyseal widening. None of these features were noted in this patient.

Conclusion

Children with Hallermann-Streiff syndrome may present with a wide range of problems requiring surgery. These include congenital cataracts, orthodontic or dental surgery, repair of cardiac defects, and antireflux procedures, as described in this infant. The postoperative course of this patient was complicated by poor healing, which has not been previously described. Fortunately the syndrome is rare but it is important to note that the anaesthetic and postoperative course can be extremely challenging.

References

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