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Anesthesia in Treacher Collins Syndrome for bone anchored hearing aid (BAHA) surgery

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Keypoints

This article describes a case of anesthesiological management in bone-anchored hearing aid (BAHA) implant placement surgery of a 7-year-old female Treacher Collins Syndrome patient. Such syndrome includes craniofacial malformations which can complicate airway management, representing a challenge for the anesthesiologist.

Abstract

Treacher Collins syndrome (TCS) is a rare genetic disorder characterized by distinctive abnormalities of the head and face, affecting about one child in every 50000. Abnormalities such micrognathia, cleft palate and cervical spine alterations are common and may represent an additional risk in airway management for general anesthesia. As a matter of fact, difficult OTI prevalence in paediatric surgery rises from 1-2% to 50% when considering the subpopulation of paediatric patients with cervical spine diseases, as TCS ones. This article describes a case report of our anesthesiological management in bone-anchored hearing aid (BAHA) implant surgery in a 7-year-old female Treacher Collins Syndrome patient born without the long process of the incus, the stapes and with hypoplasia of the oval window bilaterally. A careful preoperative evaluation and anesthesiological plan based on the use of videolaryngoscopy, routine practice in our centre, granted a successful outcome and no complications.

Keywords

Treacher Collins syndrome, pediatric difficult airway management, pediatric craniofacial malformations, pediatric videolaryngoscopy.

Introduction

Treacher Collins syndrome (TCS), also known as mandibulofacial dysostosis or Franceschetti–Zwahlen–Klein syndrome, is a rare genetic disorder characterised by dysgenesis of the hard and soft tissues of the first and second branchial arches, leading to abnormalities of head and face (Figure 1. and Figure 2.). About one child in every 50000 is affected. Features of TCS include mandibular micrognathia, conuctive hearing loss, soft and hard tissue malar deficiency, down-slanting eyes (“crying facies”), euryblepharon with or without coloboma, and cleft palate (24–28%). It can also extend to the cervical spine (17%). Oral abnormalities can lead to breathing and feeding difficulties. Brain and behavioral anomalies such as microcephaly and psychomotor delay have also been

occasionally reported as part of the syndrome. TCS is primarily caused by mutations in TCOF1 gene, but is also associated with mutations in the POLR1B, POLR1C or POLR1D genes. TCOF1 and POLR1B include an autosomal dominant heritance, while an autosomal recessive one for POLR1C.

In contrast, both autosomal dominant and recessive mutations in POLR1D have been reported in association with TCS.

Such malformations can complicate airway management under general anesthesia, increasing perioperative global risk. In paediatric surgery the incidence of difficult intubation is 0.2-5.5%, with impossible intubations of 0.08%; instead, the prevalence of difficult OTI is 1-2%, a proportion which rises to 50% when considering the subpopulation of paediatric patients with cervical spine diseases, as TCS ones.

We must not forget that 30% of anaesthesia-related accidents are caused by difficult airway management and 70% of these accidents result in permanent brain damage or even death.

This article describes a case report of our anesthesiological management in bone-anchored hearing aid (BAHA) implant surgery of a 7-year-old female Treacher Collins Syndrome patient.

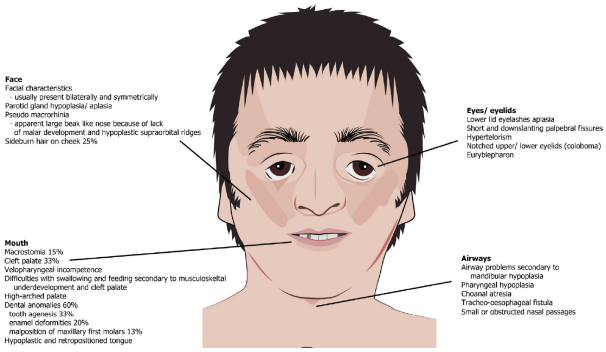

Figure 1. Typical facial features of Treacher Collins syndrome, en face view. *From Elsevier, "The surgical management of Treacher Collins syndrome". Alistair R.M. Cobb et al.*

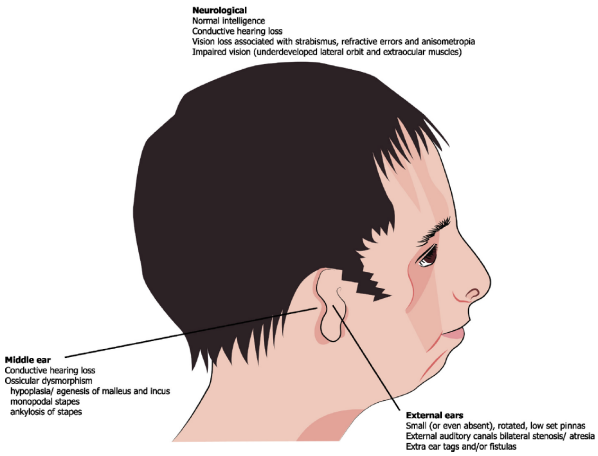

Figure 2. *"The surgical management of Treacher Collins syndrome". Alistair R.M. Cobb et al.* Typical facial features of Treacher Collins syndrome, lateral view. *From Elsevier,*

Case report and discussion

A 7-year-old Treacher Collins syndrome female patient of about 27 kg weight and 130cm height presented an ossicular chain malformation and hearing loss with indication for bone-anchored hearing aid (BAHA) implant placement surgery from the age of 2. Patient's medical history started with full-term birth from eutocic vaginal delivery after normal pregnancy, birth weight 4270g, good adaptation to extrauterine life, regular neonatal life, normal psychomotor development, vaccinations carried out according to the standard vaccination schedule. At the age of one month old, audiological screening was performed with recordings of otoacoustic emissions (TEOAE) and automatic auditory evoked potentials (A-ABR), finding a bilaterally doubtful result. Therefore, urinary CMV DNA research was carried out, which resulted negative. Auditory evoked potentials of the brain stem were performed with threshold research, with an evidence of bilateral medium degree hearing loss, more pronounced on the right side, for 2000-4000 Hz frequencies. ABR control was then programmed with threshold research by bone, resulting within the normal limits; an indication for positioning bone hearing aids was given.

One year later, on the advice of the ENT specialist, a CT ear scan was performed, showing the absence of the long process of the incus, the absence of the stapes and hypoplasia of the oval window bilaterally. An additional audiometry was performed, with an evidence of bilateral pantonal conductive hearingloss of medium degree. A first trial with a BAHA prosthesis was performed, leading to

a discreet hearing gain. An indication for BAHA implant placement surgery was then given.

Our team performed an accurated anesthesiological preoperative evaluation involving patient's parents aswell. Anesthesiological management started with the collection of an accurate familiar, physiological, pharmacological, remote and recent pathological anamnesis, which showed anything relevant apart from TCS; a detailed objective examination was also performed. Airways were evalutated according to the Colorado Pediatric Airway Score (COPUR), showing a small and moderately hypoplastic chin, an interdental space > 40mm, a partially visible uvula and an estimated head range of motion > 120°; a total score of 8 classified the difficult intubation risk as moderate. A neck circumference of 25 cm was associated with an ultrasound-measured distance from skin to epiglottis (DSE) of about 1,5 cm.

After providing a peripheral venous access (20 G at the elbow crease) and monitoring vital parameters (ECG, SpO2, NIBP, temperature by spot-on sensor, Bispectral Index with pediatric sensor, neuromuscular monitoring with TOF) and after an adequate preoxygenation, general anaesthesia was inducted with administration of fentanyl, atropine and propofol. After having verified patient's ventilation and calibration of the NMT monitoring, rocuronium was administered. Orotracheal intubation was performed using videolaryngoscopy, a routine practice in our centre, also considering the underlying pathology and the realted possible difficult intubation. Videolaryngoscopy showed a full glottic view according to the Fremantle classification (Figure 3.); 2% lidoicaine was nebulised on the vocal cords. A 5,5 sized armed endotracheal tube was placed and its positioning verified by video laryngoscopy, ETCO2 curve, pressure curve and bilateral ventilation on auscultation. Intubation was performed at first attempt, with no need for additional devices. Gastric emptying was then performed. Eyes were adequately protected with lubricating gel and atraumatic plaster. Controlled ventilation was delivered in a protective regimen (6-8 ml/kg). The maintenance of the anesthetic plan took place through the administration of sevoflurane and remifentanil continuous infusion.

Dexamethasone 0.1 mg/kg iv was given at surgery start time as a prevention of post-operative nausea and vomiting and pharyngeal edema.

Figure 3. *Videolaryngoscopic view. V*ideolaryngoscope should be use as first choice in orotracheal intubation in patiens with Treacher Collins Syndrome.

Surgical procedure started with the identification of the implantation site, 6 cm postero-superiorly from the external auditory canal. After antisepsis , preparation of the operative field and subcutaneous infiltration with local anaesthetic, a 3 cm semicircular incision centered on the implant position was performed. The flap was elevated sparing the periosteum, which was perforated over the site of implantation of the screw. A 3 mm countersink was drilled using a high-speed cutting drill. This countersink was then enlarged with a bone rasp to the shape and diameter of the implant. Finally, a 3 mm self-tapping fixture was screwed at low speed perpendicularly to the bone. The flap was placed over the fixture, sutured and perforated over the fixture with a punch scalpel, and the abutment was then screwed onto the fixture. At the end of operation, a healing cap was clipped onto the abutment, underneath which a piece of tulle gras was placed

to act as a compressive dressing. As planned, cap and sutures (Figure 4.) will be removed on the 10th postoperative day, while processor (Figure 5.) will be installed three weeks after surgery.

Paracetamol 15 mg/kg was administered at the surgical final phase, with a ward somministration every 6/8 hours. At surgey end (Figure 5.), a correct haemostasis was carefully checked. Sevoflurane and remifentanil were then stopped. Once MAC values <0.3 were reached, support ventilation was started. Extubation at the end of anesthesia was performed after Sugammadex administration with TOFr > 90%. There were no anesthesiological complications.

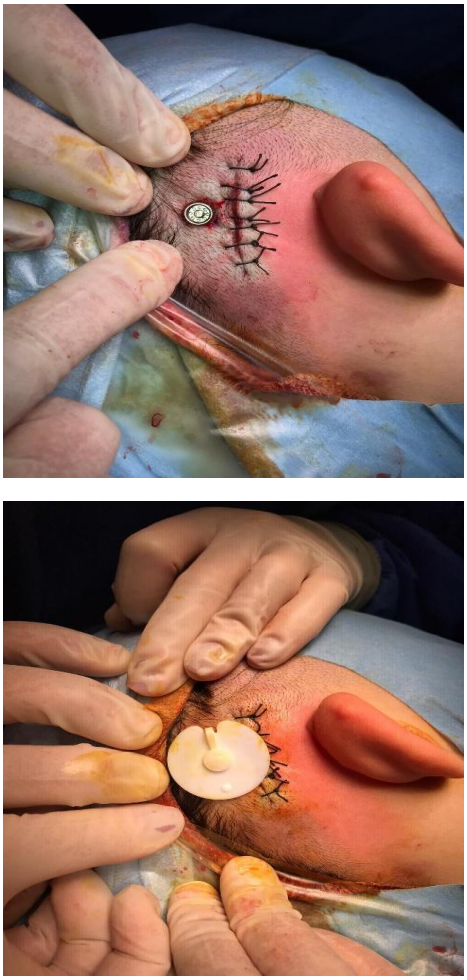

Figure 4. Bone-anchored hearing aid (BAHA) implant.

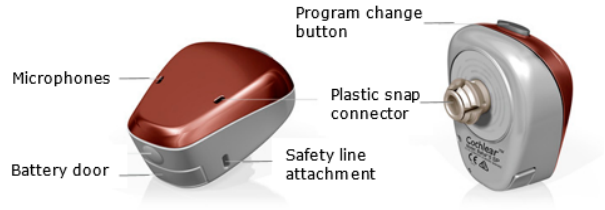

Figure 5. Bone-anchored hearing aid (BAHA) external processor. *From the Royal Victorian Eye and Ear Hospital, "Baha Recipient Information".*

An accurate post-operative monitoring has been carried out in a dedicated room adjacent to the operating block. Patient was discharged to ward after a final evalutation by ENT and anesthesiology specialists, once respiratory function, haemodynamics, state of consciousness, pain (< 4 Wong-Backer scale, < 3 NRS) and body temperature have been checked.

Conclusion

In this article, we report a case of anesthesiological management in bone-anchored hearing aid (BAHA) implant placement surgery of a 7-year-old female Treacher Collins Syndrome patient. Such syndrome includes common craniofacial malformations such micrognathia, cleft palate and cervical spine alterations which represent an additional risk in airway management for general anesthesia. As a matter of fact, difficult OTI prevalence in paediatric surgery rises from 1-2% to 50% when considering the subpopulation of paediatric patients with cervical spine diseases, as TCS ones. Videolaryngoscopy is considered the main technique to facilitate tracheal intubation and reduce its complications; its shared view promotes teamwork, reducing risks and complications related to ineffective team communication. A careful preoperative evaluation and anesthetic plan based on the use of videolaryngoscopy, routine practice in our centre, granted a successful outcome.

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