Successful management of bilateral cataracts in an adolescent subject with Down syndrome with difficult airways via surgery under modified topical anesthesia

Manejo bem-sucedido de catarata bilateral em um adolescente com síndrome de Down e dificuldade nas vias aéreas por meio de cirurgia sob anestesia tópica modificada

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ABSTRACT

We report the first case of successful bilateral cataract surgery performed under modified topical anesthesia in an adolescent with Down syndrome and a difficult airway. Airway management under general anesthesia (GA) for this patient population presents significant challenges, including increased risk of adverse intraoperative and postoperative events. This report details our innovative anesthetic approach, termed trained topical anesthesia (TTA), which facilitated uneventful successive cataract surgery in both eyes. The success of this approach highlights the importance of comprehensive psychological evaluation, patient preparation, and effective doctor–patient communication. This case report therefore proposes TTA as a viable and safe alternative to GA for cataract surgery in carefully selected patients with Down syndrome.

RESUMO

Relatamos o primeiro caso de cirurgia bilateral de catarata bem-sucedida realizada sob anestesia tópica modificada em um adolescente com síndrome de Down e via aérea difícil. O manejo da via aérea sob anestesia geral (AG) para essa população de pacientes apresenta desafios significativos, incluindo aumento do risco de eventos adversos intraoperatórios e pós-operatórios. Este relatório detalha nossa abordagem anestésica inovadora, denominada anestesia tópica treinada (ATT), que facilitou a cirurgia de catarata sucessiva e sem intercorrências em ambos os olhos. O sucesso dessa abordagem destaca a importância da avaliação psicológica abrangente, da preparação do paciente e da comunicação eficaz entre médico e paciente. Portanto, este relato de caso propõe a TTA como uma alternativa viável e segura à AG para cirurgia de catarata em pacientes com síndrome de Down cuidadosamente selecionados.

INTRODUCTION

Down syndrome (DS) is caused by triplication of chromosome 21, with an approximate prevalence of 1 in 800 live births.[1] These patients have a higher incidence of childhood cataracts than the general population. Cataract surgeries among them are usually performed under general anesthesia (GA); however, these patients are considered at high-risk for GA because of their narrow airways[2] and the presence of comorbidities such as congenital heart anomalies. The risk further increases if there is a requirement for multiple GAs, which may necessitate accommodations to limit the risks of surgery and GAs.

This work was approved by the institutional Ethics Committee of Military Hospital Wellington.

CASE REPORT

An 18-year-old male adolescent with DS was found to have severe visual impairment during a school eye health screening. Comprehensive evaluation at the eye clinic revealed that his best-corrected distance visual acuity was limited to 20/400 in the right eye (OD) and 20/200 in the left eye, with posterior subcapsular cataract in the central visual axis of both eyes (Figures 1A and 1B). Optical biometry scan (intra-ocular lens [IOL] Master 700, Carl Zeiss Meditec AG, Germany) revealed the axial lengths to be 25.02 mm OD and 24.99 mm OS with anterior chamber depth of 3.47mm OD and 3.48 mm OS. The keratometric power was derived to be K1 42.87D at 80° and K2 43.47D at 170° with astigmatism of -1.59D at 80° OD; and K1 42.44D at 88° and K2 43.71D at 178° with astigmatism of -1.27D at 88° OS. We calculated the IOL power using the SRK/T formula and aimed for emmetropia. The remaining ocular and general fundus findings were unremarkable. No comorbidities or previous surgical interventions were reported. The patient was calm, happy, and cooperative. He had a fair comprehension of verbal communication but showed mild slurring of speech. An objective evaluation of intelligence scores using the Kaufman brief intelligence test (KBIT-2)[3] revealed a nonverbal intelligence index of 64, a verbal index of 60, and a composite intelligence score of 62, indicating mild intellectual disability.[4]

The patient underwent hematological, metabolic, and coagulation panel investigations, without any reported pathological values. Given the high prevalence of cardiac abnormalities among the subjects with DS, cardiology screening was performed, which revealed unremarkable findings. Presurgical anesthetic evaluation revealed relative macroglossia and limited mouth opening, with an interincisor distance of 4 cm and a thyromental distance of

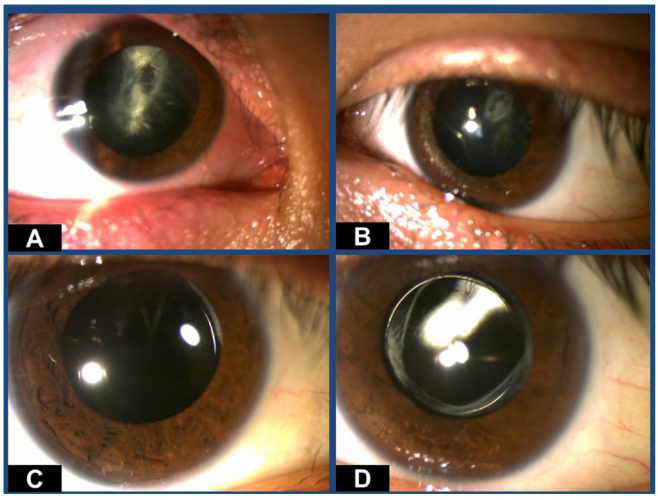

Figure 1. Clinical photographs of an adolescent with Down syndrome who underwent cataract surgery under trained topical anesthesia. Preoperative slit-lamp images revealing the presence of (A) dense posterior capsular cataract in the right eye and (B) faint posterior capsular cataract in the left eye. Postoperative slit-lamp images revealing the presence of an intraocular lens within the capsular bag (C) in the right eye and (D) in the left eye.

5.5 cm (Figures 2A to 2C). The modified Mallampati score was grade IV (only the hard palate was visible at mouth opening) (Figure 2D). He had typical craniofacial abnormalities of DS patients with limited neck mobility (STOP-BANG score of 3).[5] Considering the high risk for difficult mask ventilation and difficult intubation, the patient was classified as having a - 'class 3 physical status' according to the American Society of Anesthesiology classification.[6] The following anesthetic management strategies were initially planned: induction of GA, fiberoptic bronchoscopy-guided tracheal intubation with a single-lumen tube and preparation for rescue wire-guided cricothyroidotomy for cannot intubate/cannot ventilate (CICV) situations.

Considering these factors and avoiding the high risk involved in administering GA twice to the same patient in different settings, we planned to perform cataract surgery under conventional topical anesthesia with a simple modification to ensure additional safety, with the GA being kept as a backup.

Initially, rapport was established with the subject adolescent and his parents through frequent friendly interactions during clinical assessment. The ability to endure topical anesthesia was subsequently assessed by analyzing the patient's response to light touch and pressure in a simulated conducive surgical environment created in the minor operating theater (OT) of the department. His response and cooperation to follow the verbal directions, ability to lie still, be relatively flat, tolerate the face being covered by a drape, hold the head still, and focus

Figure 2. Preanesthetic assessment revealing (A) a relative macroglossia, (B) an interincisor distance of 4 cm (less than 3 finger depths), (C) a Grade II hyo-mental distance of 5.5 cm (indicated by the blue arrow) and (D) a Grade IV modified Mallampati score (only the hard palate was visible at mouth opening).

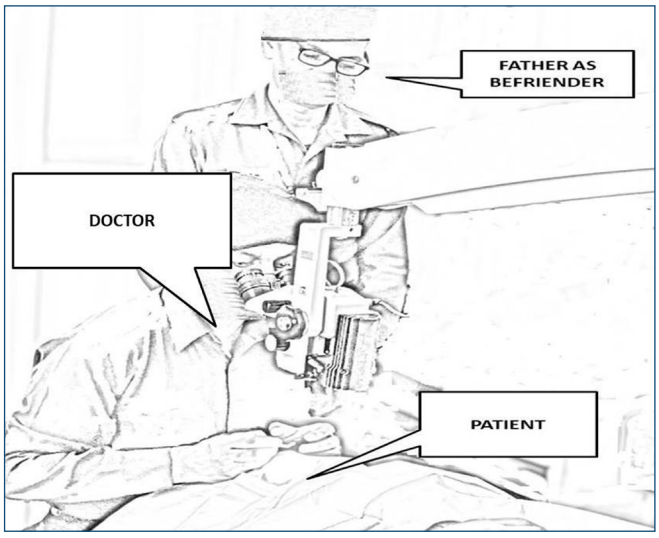

Figure 3. A representative illustration of the trained topical anesthesia, wherein an adolescent with Down syndrome is assessed preoperatively under topical anesthesia for his response to the light touch and light pressure, following the verbal directions of the doctor, ability to lie still and relatively flat, ability to tolerate their face being covered by a drape, ability to hold the head still, and focus constantly on the light emanating from the microscope, in a simulated operative environment, in the presence of a befriender as his father.

constantly on the light emanating from the microscope were assessed subjectively and were found to be satisfactory (Figure 3). He was accustomed to the simulated surgical environment by putting on a surgical drape and eye speculum seven days before and one day before surgery. Additionally, his father was kept as a 'befriender' for emotional support. Pep talks, with the help of his parents, were used when necessary to make him comfortable. His father was scheduled for a constant bystander during the preoperative procedures and live surgeries in the OT. This simple modification of topical anesthesia was termed 'trained topical anesthesia' (TTA). Written informed consent was obtained from the patient's father before surgery.

Each eye was planned and operated as a single separate surgery. On the day of first eye (OD) surgery, the patient appeared calm and cooperative (Richmond Agitation Sedation Scale score of +0). A team of anesthesiologists monitored the patient and was well prepared to administer GA if needed. In cases of untoward complications or circumstances, the backup plan was to temporarily halt the on-going surgery, patch the eye, and perform corrective surgery under GA, for which prior additional high-risk consent was obtained from the patient's parents.

Proparacaine (0.5%) was instilled in the eye to be operated on 5 min before surgery and immediately before creating the first corneal incision for achieving anesthesia. After cleaning the lid and periorbital skin with 5% povidone-iodide solution and draping, a self-retaining Barraquer wire eyelid speculum was inserted to expose the globe. Povidone-iodine (5%) was applied for 2 min over the exposed area, followed by flushing out with sterile irrigating solution. Two paracenteses were created with 15-degree safety sideport knives (BVI®) in the OD at 11 o'clock and 7 o'clock, leaving 9 o'clock for the main incision, with a keratome of 2.4 mm (BVI®), which was made later for the phacoaspiration and IOL insertion. The 1 and 5 o'clock positions were used in the left eye, leaving the 3 o'clock position for the main incision. We used trypan blue (0.06%) for better visualization of the anterior lens capsule and hydroxypropyl methyl cellulose (2%) as ophthalmic viscoelastic devices (OVDs) to maintain the space. Following hydro-dissection with a 27-gauge cannula, phacoemulsification of the nucleus was carried out using 'stop and chop' technique under a surgical microscope (OPMI Lumera 700, Carl Zeiss Meditec AG) and the Stellaris Elite phacoemulsification system (Bausch & Lomb, Rochester, NY, USA) with almost minimal requirement of emulsification energy. We used foldable acrylic aspheric hydrophobic UV-blocking monofocal posterior chamber (Tecnis Eyhance IOL, Johnson & Johnson Vision, Santa Ana, CA USA), which, being non-preloaded, was manually loaded into a compatible injector and inserted through the original main incision using wound-assisted delivery. Viscoelastic was removed with bimanual irrigation/aspiration hand-pieces through the paracenteses.

The incision was sealed by creating incisional edema. No augmentation of topical anesthesia was required during surgery. The postoperative therapeutic regimen was topical antibiotic with corticosteroid (Moxifloxacin 0.5% with Dexamethasone 0.1%, Vigadexa®, Novartis) drops with a decreasing dosage. The follow-up visits were the next day, 1 week, 1 month, 3 months, 6 months, and 1 year. Visual acuity, autorefraction, tonometry, slit lamp examination and posterior segment OCT were performed at all visits.

At the 7-day postoperative visit, the patient was delighted with their improved vision and expressed his desire to get his other eye operated on at the earliest. At the 1-month postoperative visit, refraction was performed in the first eye (OD) to adjust the calculation for the second eye IOL. Uneventful cataract surgery with similar protocol was successfully performed with this technique in left eye after a gap of one month (Figures 1C and 1D). The drape time, as calculated from covering the face to removing the ocular drape was 22 min for the OD and 20 minutes for the left eye. The surgical time, as calculated from the time of making the first corneal incision to the completion of hydration of the incision ports, was 15 min for the first eye and 12 min for the second eye, which was within the range of surgical time (10 to 16 minutes) taken by the operating surgeon for the completion of cataract surgery of similar grade in routine patients. Final refraction was performed in both eyes 12 weeks after the second eye was operated, which was +0.50 DS/-1.0 DC at 70° OD and -0.25 DS/-0.75 DC at 110°OS, with a final best corrected distant visual acuity of 20/40 OD and 20/20 OS. Post-surgical uncorrected near visual acuity was N24 on Snellen's near vision chart in both eyes, increasing to N6 with a reading addition of +2.00 DS for a near distance of 40 cm. In the follow-up one year after surgery, the eyes remained quiet, visual acuity and intra-ocular pressure were 14 mmHg in both eyes as measured by a non-contact tonometer (Topcon Corporation, Tokyo, Japan).

DISCUSSION

Although topical anesthesia is not the anesthesia of choice for persons with mental impairment, subjects with DS have some special characteristics. They usually have cheerful personalities and are socially amicable. They were found to have a calm mental status and obey directions, as did regular patients.[7] The average IQ of these individuals is usually greater than 50, despite their cognitive impairment.[8] While children with DS do not have as deep a vocabulary as their typically developing peers do, their receptive vocabulary can be seen as a relative asset.[9] Their deliberate use of social gestures and language remains on par with their mental age, despite their weaknesses in expressive phonology.[8,9]

Nevertheless, the basic essence of TTA is the creation of a stress-free environment in the OT, by which the patient can feel relaxed but still be able to communicate.[10,11] One way to achieve this is through constant reassurance or pep talks by the operating surgeon[10] or a befriender.[12] Parents can be most suitable befrienders in such cases and can additionally provide tactile reassurance by holding their hands throughout the procedure. The role of the handholder has recently gained importance, as the amount of local anesthetic administered has decreased.[12] Additionally, as the patient in our case was required to remain alert and actively participate in 'surgeon-patient-befriender' communication, preoperative anxiolytics were not used, as most of them have been reported to impair psychomotor performance as well as immediate and delayed recall and recognition.

While TTA in our patient appeared to be safe and comfortable, it required a cooperative patient as well as a calm and experienced surgeon. This is likely driven by surgical expertise, comfort levels, and the complexity of the procedure. TTA is not applicable to surgeons who prefer no eye movement or communication during surgery. The speed at which the surgeon operates is also a consideration. In addition, a thorough history evaluation and establishing a rapport with the patient with empathy are necessary.[10] TTA may also not be suitable for subjects under 15 years of age, and procedures which are expected to take longer than 20 minutes or complex cases.

To the best of our knowledge, this is the first report describing cataract surgery under topical anesthesia with some modifications in a case of DS. A preplanned management strategy, awareness of the subject's cognitive levels, simulated training, pep talk, establishing rapport with the patient, and the use of parents as befrienders may help adolescent DS subjects with challenging airway cases undergo cataract surgery under TTA and should be explored as alternatives to the GA whenever feasible.

AUTHOR'S CONTRIBUTION

Rakesh Kumar Jha developed the conception and design of the study and was in charge of the analysis and interpretation of results and writing the first draft of the manuscript. Bhupesh Bhatkoti was in charge of the interpretation of results and critical review of the final manuscript. All authors have approved the final version of the manuscript and are responsible for all aspects of it, including ensuring its accuracy and integrity.

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