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Welcome to the first edition of 2025, and the start of Volume 31.

This quarter's edition has three papers, so an improvement on the last issue, but I would still welcome your dedication to this publication with a continuation of submissions to the world's premier Journal on all aspects of Ambulatory Surgery. This Journal would surely fail without your continuing input.

The papers in this edition include a study evaluating the role of music as an intra-operative adjunct, a case report describing "Empty Nose Syndrome" and its management in an outpatient setting, and the care of a patient with myotonia congenita undergoing ulnar nerve decompression in a daycase setting.

Bates and colleagues report on a study evaluating the choice of music provision during cataract surgery in an English hospital, where they compared the preferred music choice of patients. Predictably perhaps for the patient cohort, classical music was judged as the most popular request. However, the authors also commented on the overall benefit not only of music choice as a patient empowerment, but also as a beneficial emotional adjunct that as a method of distraction, is simple, effective and inexpensive.

An American case report describes the management of a rare otolaryngological condition, where despite previous turbinate surgery, patients still experience a paradoxical nasal obstruction despite an objectively wide and patent nasal fossa. The aetiology of this condition is poorly understood, with a potential two cause process affecting both damaged tissue and damaged sensory neural input, potentially leading to psychological symptoms of anxiety depression or suicidal ideations. The report describes the successful surgical option of using acellular dermal sheets to reconstruct pseudo-inferior turbinates, and the importance of multidisciplinary collaboration that is possible in an ambulatory environment.

The final paper is an Irish case report of ulnar nerve compression in a patient with myotonia congenita. The pathophysiology of the disease is comprehensively described and the methods by which symptoms can be avoided are discussed.

In conclusion, keep those submissions coming to ensure the next publication date in June.

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Trends in music preferences of patients undergoing routine cataract surgery

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Abstract

This study examined music preferences among 272 patients undergoing awake cataract surgery at the Wye Clinic over eight months. Patients were asked about their preferred music, and responses were categorised with input from professional musicians. Classical music was the most requested genre (28.7%), followed by “anything” (11%), jazz (8.5%), pop (7.7%), and

Keywords: Cataract Surgery, Music, Patient choice.

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rock (7.4%). Results suggest most patients appreciate the opportunity to choose music, enabling a sense of control and enhancing their surgical experience. These findings support incorporating patient-selected music into surgical care to improve comfort and satisfaction.

Introduction

Music therapy is “the professional use of music and its elements as an intervention in (a) medical... environment with individuals... who seek to optimize their quality of life...” [1]. In the absence of a qualified music therapist, the music used as a passive environmental adjunct during surgery is considered a form of receptive music therapy [2].

When used as a complement to the operative journey, music consistently demonstrates a matrix of positive effects. Psychologically, there are associated reductions in perioperative and postoperative levels of subjective pain and psychological distress measured using the Hospital Anxiety and Depression Scale (HADS) and the EuroQol 5-dimension 3-level test (EQ-5D-3L) [3,4]. There may be a counteraction of the feelings of helplessness and hopelessness that come with the emotional burden of being in hospital [5]. Physiologically, patients benefit from an associated reduction in systolic blood pressure and heart rate, which has been reported within an ophthalmic surgical setting, among other surgical settings, including elective gynaecological surgery [3,6,7,8,9]. These effects exist for elderly patients, and are not significantly affected by medically treated hypertension [10]. In particular, patients who tend towards hypertension during acute stress may benefit more from music therapy in a surgical setting than normotensive patients [11].

An array of positive effects is demonstrated from using music as a complement to ophthalmological procedures. For example, playing Mozart to patients immediately prior to automated perimetry is associated with reductions in fixation losses, false positives and negatives, and test duration [12]. There is an associated reduction with the surgical challenges of head drift and eye movement during awake cataract surgery [13]. Despite the large body of literature examining the value of music within surgery, a relatively small number of papers investigate its value in ophthalmic procedures. This work aims to identify what music patients undergoing awake cataract surgery actually want to listen to. As the use of a general anaesthetic is now uncommon practice in routine cataract surgery, the value of non-invasive measures such as music, with the potential for reductions in the psychophysiological stress evoked by having surgery, hold importance.

Methods

Data was collected from 272 consecutive routine cataract (phacoemulsification with intraocular lens implantation) surgeries (all under topical anaesthesia) over an eight-month period, using a simple form (see Supplementary information - .docx file). This collected simple anonymised patient demographic data, alongside expressed music preferences after being asked the open question of what patients would like to listen to for the duration of their cataract surgery. Responses were initially grouped into five broad categories:

1. ‘Specific genre’
2. ‘Specific era’
3. ‘Other’
4. ‘Surgeon’s preference’
5. ‘Wanted no music’

After data collection, group frequencies were recorded, and the ‘Other’ category was sub-grouped into genres. The advice of two professional musicians was sought at this stage. Descriptive data analysis was conducted in R (4.2.3, packages: ggplot2, dplyr, broom and stringr) [14, 15, 16, 17, 18]. Note, NHS REC was not required, as this project is not research according to the NHS HRA decision tool.

Results

In total, data relating to patients’ music choices during 272 routine phacoemulsification procedures were collected. Patient ages followed a normal distribution between the range of 34-94 years with mean of 74.9 (8.9). There were 147 females and 105 males (20 had no recorded sex).

During the eight months of data collection, 265 of 272 (97.4%) of patients undergoing routine cataract surgery did want to listen to music during their time in theatre. Of the 7 (2.6%) who did not want to listen to music, two were deaf. Of the 265, 117 (43.0%) made a specific request, which referred to a specific instrument/ artist/ song, etc. Specific requests were subcategorised by genre, and a comprehensive breakdown is shown in Table 1. 88 (32.4%) requested a specific genre, and these are shown in Table 2. 37 (13.6%) made vague requests, which included the statements shown in Table 3. 17 (6.3%) asked for the surgeon’s preference to be played, and seven

Table 1 Genres of specific requests (117 requests in total).

Genre	Count (% of 117)
Pop	19 (16.2)
Rock	17 (14.5)
Baroque	13 (11.1)
Classical	13 (11.1)
Easy listening	12 (10.3)
Mainstream jazz	9 (7.7)
Romantic modern	9 (7.7)
Romantic	7 (6.0)
Folk	5 (4.3)
Soul	5 (4.3)
Blues	5 (4.3)
Instrumental	4 (3.4)
Classical romantic	3 (2.6)
Modern	3 (2.6)
Big band	3 (2.6)
Traditional jazz	2 (1.7)
Jazz fusion	2 (1.7)
Country	2 (1.7)
World	2 (1.7)
Classical opera	1 (0.9)
Choir	1 (0.9)
Bebop jazz	1 (0.9)
Modern jazz	1 (0.9)
Rhythm and blues	1 (0.9)
Progressive rock	1 (0.9)
Dance	1 (0.9)
Electronic/ trance	1 (0.9)

Table 1 shows the raw counts and associated proportions for the genres of different specific requests, made by those in the sample who made specific requests.

(2.6%) requested a particular era of music, which is shown in Table 4. Where there are overlaps between how patients' requests were grouped, this data is combined in Table 5 (overlapping subcategories are italicised in Tables 1-4).

Discussion

The data suggest that the most patients undergoing routine awake cataract surgery do want to listen to music while in theatre. The top five most popular requests were classical music (28.7% of the total sample), 'anything' (11%), jazz (8.5%), pop (7.7%) and rock (7.4%). The categorisation of genres was conducted with two professional musicians of different backgrounds (romantic-modern, and jazz). There is a general tendency to group together similar genres with classical music. For example, baroque, modern and more broadly, instrumental, may be grouped together erroneously under the heading of classical. Similarly, jazz artists from the requests were broken down into five distinct sub-genres: mainstream jazz, traditional jazz, bebop jazz, modern jazz and jazz fusion. Of note, two

Table 2 Requests for genres (88 requests in total).

Genre	Count (% of 88), where relevant, sub-requests with associated counts are specified
Classical	65 (73.9) Gentle 14 Light 3 Piano 2 Relaxing 1 Soothing 1
Jazz	8 (9.1) Soft 2 Traditional 1
Country, or Country and Western	3 (3.4) American 1
Rock	2 (2.3) Hard 1
Baroque	2 (2.3)
Easy listening	2 (2.3)
Opera	1 (1.1)
Doo-wop	1 (1.1)
Elevator music	1 (1.1)
Military music	1 (1.1)
Pop	1 (1.1)
Reiki music	1 (1.1)
Soul	1 (1.1)

Table 2 shows the raw counts and associated proportions for different genres, from those in the sample who requested specific genres.

Table 3 Vague requests (37 requests in total).

Vague request	Count (% of 37)
Anything	30 (81.1)
Something relaxing	2 (5.4)
Gentle, not rock	1 (2.7)
As little as possible	1 (2.7)
Anything, older stuff	1 (2.7)
Anything relaxing	1 (2.7)
Anything but jazz or heavy metal	1 (2.7)

Table 3 shows the raw counts and associated proportions for different vague statements, from those in the sample who requested music vaguely.

Table 4 Requests for particular eras (7 requests in total).

Era	Count (% of 7)
60s	4 (57.1)
80s	2 (28.6)
90s	1 (14.3)

Table 4 shows the raw counts and associated proportions for different eras, from those in the sample who requested music by specifying an era.

Table 5 Combined data for overlapping categories.

Overlapping genre	Count (% of total sample, 272)
Classical	78 (28.7)
Jazz (including mainstream jazz, traditional jazz, jazz fusion, bebop jazz and modern jazz)	23 (8.5)
Pop	21 (7.7)
Rock (including progressive rock)	20 (7.4)
Baroque	15 (5.5)
Easy listening	14 (5.1)
Soul	6 (2.2)
Country, or Country and Western	5 (1.8)
Opera (including classical opera)	2 (0.7)

Table 5 shows the raw counts and associated proportions of the total sample for sub-categories that were overlapping in other groups (which can be identified in Tables 1-4).

patients requested Classic FM, which were not included in the overall 'classical' genre, as Classic FM plays a range of genres. Also note that calculated total counts from the tables may be higher than expected, as some requests fitted into more than one genre, and a small minority of patients requested more than one category, e.g. '90s pop'. Specific requests were classified by genre (Table 1), because this was the most useful approach for data integration (Table 5). Alternative approaches to music categorisation could focus on specific musical elements such as the tempo of the songs played, or the instrumental/vocal focus, etc., though the data collected here does not facilitate this type of evaluation.

This study deliberately does not focus on any anxiolytic or other psychophysiological effect that music has on cataract surgery patients. This is well-established already, and patients at the Wye Clinic often receive diazepam prior to surgery, so there would be low power in this setting to examine any anxiolytic effect of music. The sample size of 272 cataract surgeries, across the time-span of eight months suggests high internal validity of painting a reasonable picture of what music choices patients make when undergoing routine awake cataract surgery. This data was collected from a single private clinic, and generalisability to different geographical locations should be considered carefully.

The effect of choice

The importance of being specific with the categorisation of patients' music choices arises when considering the array of effects that music may have on them. Though on the surface, the perceived effect of highly different requests, such as progressive rock compared to light classical may be contrasting, Hole et al reported no significant differences between genres on outcomes including post-operative pain, analgesia use or length of stay after surgery [4]. Interestingly, they also found no significant differences between patients having the choice of music played versus not having the choice. In contrast, Tan et al found that in a surgical setting, patients generally prefer to make their own choices of music [3]. This is supported by the data in this paper. Patients' mental states, personality traits and individual temperaments may be important factors that influence their psychophysiological responses to music [19]. The psychological elements implicated in music's positive role in elective awake

surgery include an internalising of locus of control, and self-efficacy through playing an active role in the imposition of a more relaxed physiological state [20,21]. Drawing this together, the actual choice of music may not be as important as the patient making the choice, given that genres that evoke a relaxed physiological state are likely to differ at least slightly between individuals.

Emotional state

Though much of the literature in this area present correlations between music and positive effects seen in patients as causal, listening to music is associated with limbic system alterations [22,23]. Stefano et al, found a statistically significant decrease in IL-6 (proinflammatory), and suggested that more plasma morphine was converted to morphine-6-glucuronide, indicating a potential physiological basis for decreased blood pressure from listening to music [24]. Endogenous opiates (endorphins) are likely to be involved in the psychophysiological euphorogenic effects of music [25]. Furthermore, individuals listening to music specifically chosen for pleasure is accompanied by a measurable increase in dopaminergic activity within the mesolimbic reward system [26].

Additionally, with lower order processing, the stimuli provided by musical elements elicit psychophysiological responses rooted in enhancing or minimising states of arousal, including responses in arousal state to concurrent stimuli, such as the possible perceived threats in an operating environment. Beyond this, the contextual associations with music – for example, with different genres – that arise from higher order processing, interact with how these psychophysiological responses occur. For a patient, the experience of awake surgery is likely to be a sequence of somewhat unexpected events. The deliberate use of music may provide an auditory object that is predictable to the patient, particularly if the song, artist or genre was chosen by them because of their familiarity with it. In doing so, the heightened sense of arousal that accompanies unpredictability is dampened [27,28].

Literature on the use of music in an operating environment is limited in its applicability to ophthalmic procedures. Indeed, neither Bernatzky et al's review of the therapeutic potential of music in healthcare, or Hole et al's systematic review and meta-analysis addressing the value of music for post-operative recovery examined any studies that focused on ophthalmic surgeries [2,4]. This paper acknowledges that extensive literature has been done to show what value music has for the patient undergoing awake surgery, and so begins to examine what music specifically carries the most value. Future enquiry into this may include asking patients what music they would like to listen to during pre-operative counselling, as asking on the day of surgery may make some feel as though they are being put on the spot with unfamiliar people, i.e. the theatre team. This may be a confounding factor that disguises the true music choices patients wish to make, and perhaps is more significant for those with a high external locus of control. In the same vein, another approach might be to ask patients to write their choice down, instead of verbalising it. This action may reduce the possible perception of being judged for personal musical choices.

Another important question is whether or not music has an effect on surgical skill. In a Gedanken experiment aiming to measure the highest possible surgical outcomes by somehow incorporating music into the theatre environment, there are two conditions to examine. On one hand, if the patient's ability to remain calm and still are maximised, their outcomes are likely to be better than if they were anxious and agitated. On the other, if the surgeon's ability to achieve and maintain a calm and focused state are enhanced, patient outcomes will also be better. In a group of 14 ophthalmologists and 12 ophthalmology residents, listening to Mozart while using the EyeSi surgical simulator, the roundness of capsulorrhexis (a key step

in cataract surgery) improved by 33.1% ($p=0.0367$) and total scores by 23.3% ($p=0.0249$). Surgical skill was not found to be a significant confounding variable [29]. However, if a statistically significant difference exists between the surgical outcomes when the focus is on optimising patients' relaxation and stillness, or surgeons' calmness and focus, it is likely to be lost in a blurry confidence interval. Clearly, optimising both would be ideal. Headphones under the surgical drape could be considered where there are differences in musical taste between the patient and the operating surgeon [10], although this should be considered against the reduced ability of the patient to listen to instructions.

Conclusions

Music offers a reliable, controllable object to patients undergoing awake day surgeries – here, cataract surgery. Most do want to listen to music, and will make a choice given the chance. As a method of distraction, it is simple, effective, and inexpensive. This paper describes that classical music is the main preference among patients undergoing routine awake cataract surgery.

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Optimizing Anesthetic Management of Empty Nose Syndrome at an Ambulatory Surgery Center

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Abstract

Aims: This report explores a case of Empty Nose Syndrome (ENS) following turbinate reduction surgery, focusing on management strategies in an outpatient setting.

Methods: We present a case of a young male with ENS and obstructive sleep apnea who underwent surgery using acellular dermal sheets to reconstruct pseudo-inferior turbinates. A multidisciplinary approach involving ENT, anesthesia, and nursing teams was utilized.

Results: The surgery was successfully performed on an outpatient basis, demonstrating the feasibility of managing ENS with careful planning.

Keywords: Empty Nose Syndrome, turbinate reduction, outpatient surgery, multidisciplinary approach.

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Conclusion: This case highlights the importance of multidisciplinary collaboration and the need for further research to optimize ENS treatment strategies.

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Introduction

Empty Nose Syndrome (ENS) is a rare but debilitating condition that occurs following surgical procedures such as turbinate reduction or nasal surgeries. Despite the nasal passages appearing anatomically patent, patients experience paradoxical nasal obstruction and chronic nasal dryness, resulting in significant psychological distress. The pathophysiology of ENS remains poorly understood, making its diagnosis and management challenging [1–3]. This can lead to misdiagnosis, often those that are psychosomatic.

Dr. Steven Houser defines ENS as “paradoxical nasal obstruction despite an objectively wide, patent nasal fossa.” Patients commonly report a “stuffy” or “empty” feeling, due to impaired airflow sensation, often following total or partial turbinectomy. ENS cases consistently involve some form of turbinate surgery [4].

ENS patients may experience nasal crusting, thick mucus, and occasional pain. Some report improved breathing during upper respiratory infections, adding to the complexity of the condition [3].

ENS is often mistakenly identified as synonymous with atrophic rhinitis in the literature. While they share symptoms such as congestion, dryness, and crusting, they are distinct conditions with different origins. Atrophic rhinitis, which can be primary or secondary with various underlying causes, involves the loss of turbinate and mucosal tissue. In contrast, ENS is an iatrogenic condition resulting from medical intervention. This distinction is crucial because secondary atrophic rhinitis can follow turbinate surgery or arise from trauma, infection, or immune issues, and it has known pathogens, unlike ENS, which does not have a specific associated pathogen [5].

It is still unclear as to why some patients will develop ENS and others will not. One theory is a “two hit” process that can occur. First, tissue is excised or damaged, and second, the sensory nerves to the affected area regenerate poorly. The meatuses that are formed between the turbinates provide resistance to airflow and create a laminar flow

pattern. Airflow is disrupted after procedures such as a turbinectomy which destroys the relationship between the turbinates and meatal structures. The most common combination symptoms used to diagnose ENS are paradoxical airway obstruction, dyspnea, nasal and pharyngeal dryness, hyposmia, and depression [3].

The development of ENS is unpredictable; patients with complete turbinate removal may never develop the syndrome, while others with only partial resection experience significant symptoms. Symptom severity and impact on quality of life vary, with some patients experiencing a constant sense of suffocation. Depression and reduced smell acuity are also common. ENS can manifest months or even years after surgery, often due to continued tissue atrophy. The loss of nasal mucosa appears to be a key factor, leading to altered airflow and deficient sensation [2].

Traditional diagnostic tests, like rhinomanometry and acoustic rhinometry, often fail to align with the patient’s subjective experience of obstruction. These tests measure airflow resistance and nasal cavity dimensions but do not fully capture the neurosensory aspects that influence nasal patency perception. Studies show that trigeminal afferents play a significant role in this perception; for instance, local anesthetics can induce a sensation of obstruction, while menthol can create a feeling of decongestion without actual changes in nasal structure [3].

Anesthesia can be challenging in patients who present for ENS repair, especially in a fast-paced outpatient surgery center. Foremost, it is imperative that anesthesiologists recognize the profound emotional and physical toll that many ENS patients endure. When these patients present for surgery, they are often frustrated and anxious as they may have already undergone multiple procedures without relief. Their fear is rooted in the debilitating symptoms of ENS, and it is vital for anesthesiologists to approach them with empathy, understanding the complexities of their condition and the profound impact it has on their quality of life. Patients are often plagued with psychological symptoms including anxiety, depression, and in extreme cases suicidal ideations. Patients are often on a myriad of medications

pre-operatively which can make the administration of anesthesia challenging in an outpatient surgical center.

As ENS is a rare condition, there are no clear anesthetic guidelines for managing ENS patients undergoing surgery. Given the complexity of ENS our case presentation underscores the importance of a multidisciplinary approach in managing this condition.

Methods

The patient was a male in his early 20s, with a BMI within the normal range. The patient had a past medical history of obstructive sleep apnea, and a condition related to nasal function. His surgical history included multiple prior nasal surgeries, including turbinate reductions performed in a different country. The patient traveled a significant distance to present to the ambulatory surgery center for a procedure involving the placement of acellular dermal sheets to create pseudo-inferior turbinates, aiming to alleviate his symptoms and improve nasal function.

Results

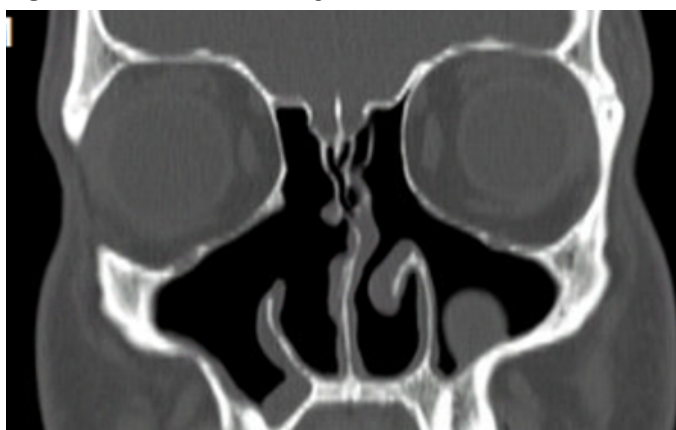
Preoperative

His ENT consultation was notable for dry nasal mucosa with small inferior turbinates. An endoscopic evaluation showed a reasonably straight septum, dry mucosa, no polyps, and open maxillary sinuses with significant turbinate loss. The cotton test, simulating a submucosal implant, provided approximately 70% improvement in breathing. The patient's outside hospital computed tomography (CT) sinus data was reviewed (Figures 1, 2).

Figure 1 Coronal CT scan image of the nasal anatomy. Coronal CT view showing a noticeable absence or reduction in the turbinate structures.



Figure 2 Coronal CT scan image of the nose and turbinates



In preoperative holding, his vital signs were stable though he appeared anxious. He was verbally reassured and given time with his family and support persons prior to premedication with midazolam and was then brought to the operating room.

Intraoperative

The patient was preoxygenated with the anesthesia mask and 10 liters of oxygen near, not on, his face due to being intolerable to the feeling of the oxygen flow. General anesthesia induction included fentanyl, propofol, lidocaine, and rocuronium and an endotracheal tube was placed with a Mac 3 blade. General anesthesia was maintained with a remifentanyl infusion and sevoflurane. Dexamethasone and cefazolin were also given, along with a dose of ondansetron before awakening. At the conclusion of the surgery, the patient had a smooth emergence and was taken to the recovery room.

A coronal section which shows the inferior turbinates that are significantly reduced, creating a more open nasal passageway.

Postoperative

Postoperatively the patient's vital signs were stable, including pain rated at 3/10 and no nausea nor vomiting. Due to unanticipated concerns from the support persons, the PACU course was prolonged to nearly three hours.

Discussion

ENS presents a significant challenge for both patients and healthcare providers due to its complex symptomatology, considerable psychological challenges, and impact on quality of life [1, 2]. This condition can be particularly demanding for anesthesiologists. To address the complexities of ENS, it is crucial to adopt a multidisciplinary approach. This involves coordination between anesthesiology, ENT surgery, and nursing teams to ensure comprehensive management and improve postoperative outcomes [1].

During surgery, anesthesiologists should be ready for possible challenges in managing the airway due to nasal obstruction or altered anatomy. Using flexible fiberoptic bronchoscopy or video laryngoscopy can be particularly helpful in these situations. Ensuring proper hydration and humidification of the inhaled gases is helpful to prevent further nasal dryness and crusting, which are common in patients with ENS. The selection of anesthetic agents should focus on creating a motionless surgical field for operation while also planning for postoperative pain relief and minimizing respiratory depression [6]. Local anesthetic and intravenous opioid medication were given intraoperatively by the surgeon and anesthesiology, respectively.

After surgery, anesthesia—focus should be on effective pain management, nasal hygiene, and emotional support. Using a mix of pain relief methods, including local anesthetics and non-opioid medications, helps manage pain while keeping respiratory function intact. Regular nasal saline rinses and humidification are essential for maintaining moist mucosa and preventing dryness, which is crucial for patient comfort and optimal surgical outcomes. It is vital to work closely with nursing staff for ongoing monitoring and support [7].

Preoperative planning should include contributions from all relevant specialties to create a personalized care plan for the patient including considerations for lodging, meals, post-surgical care, follow up appointments, and other social and mental health support. Ongoing communication and teamwork among all involved can help tackle any challenges that come up during the perioperative period and enhance overall patient outcomes [2,7].

This case adds to the literature by demonstrating that a patient with ENS can be managed in an outpatient setting without complications, provided that a thorough evaluation is conducted, and the team adapts to the patient's needs.

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Ulnar Nerve Decompression in a Patient with Myotonia Congenita under Regional Anaesthesia in a Daycase Setting: A Case Report

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Abstract

Myotonia congenita is a rare channelopathy characterized by impaired skeletal muscle relaxation after a depolarization causing myotonia. Several perioperative factors can trigger myotonia which can result in life-threatening muscle spasms with resultant ventilation difficulties, hence majority of these patients are not considered for ambulatory surgery. There is also a gap in the literature surrounding the application

of peripheral nerve blockade in this patient cohort. We report the case of a patient with Myotonia congenita who successfully underwent ulnar nerve decompression in a day-case setting under regional anaesthesia and sedation highlighting the importance of proper patient selection and preparation.

Keywords: Myotonia congenita, day case surgery, ambulatory anaesthesia, regional anaesthesia.

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Introduction

Myotonia congenita (MC) is a rare genetic disorder affecting skeletal muscle excitability, with a worldwide prevalence of about 1:100,000 (1). MC is caused by a mutation in the muscle chloride voltage-gated channel 1 gene (CLCN1) on chromosome 7q35. The clinical manifestations can vary widely, from subclinical symptoms to severe myotonic reactions and muscular weakness.

Neuromuscular disorders are rare but can have serious implications for these patients when undergoing surgery under anaesthesia (2). Association with musculoskeletal, respiratory, cardiovascular, gastrointestinal, metabolic, and central nervous system disorders warrants careful perioperative planning to achieve the best outcomes for this complex patient cohort. The anaesthetic management of a patient with MC should involve a careful emphasis on minimizing myotonic reactions, which can be triggered by several precipitants like, hypothermia induced shivering, pain (including pain on propofol injection), emotional stress, prolonged fasting and drugs such as depolarizing muscle relaxants (e.g., suxamethonium), adrenaline, beta-adrenergic agonists, and propranolol.

Most patients with MC are not considered for ambulatory surgery given the associated perioperative challenges. To our knowledge, there have been no published case reports of ambulatory anaesthesia as an option for managing patients with MC. In addition, there is a gap in the literature surrounding the efficacy of peripheral nerve blockade in this patient cohort. Our case report demonstrates that with proper patient selection and careful preparation successful ambulatory care under regional anaesthesia (RA) is possible for patients with MC. Written informed consent was obtained from the patient and the manuscript adheres to the applicable EQUATOR guidelines.

Case description:

A 36-year-old female required decompression and transposition of the ulnar nerve. She was a known case of MC (Thomsen's type 1) since age 30, with a mutation in the CLCN1 gene with co-expression of sequence variant c.811T>C:P.Cys271Arg (reported as being consistent with but not diagnostic of autosomal dominant {AD}

inheritance). Electromyography and nerve conduction studies had been performed to confirm the diagnosis of MC, and a muscle biopsy had excluded muscular dystrophy.

She first noticed symptoms of stiffness aged 8, predominantly affecting her upper limbs, which was precipitated by hunger, stress, cold temperature, and infections. Worsening of myotonia was observed during her pregnancy 6 years ago when she had difficulty standing from seated. She reported stiffness in the neck which improved on movement, keeping with the warm-up phenomenon which is a well-known attribute of the disease. The patient was prescribed both phenytoin and mexiletine in the past, but therapy was discontinued due to the limited benefit.

Before undergoing this surgery, the patient was reviewed by the consultant anaesthetist assigned to the case. Following a multidisciplinary consultation with the surgical team regarding the duration of the procedure and anticipated postoperative complications, a plan was made to proceed under RA and propofol based sedation in a day case setting.

Preoperatively the patient was advised to minimize fasting time to 6 hours for solids and 2 hours for clear fluids as hunger had been noted as a precipitant of her myotonia. She was listed to be the first case on the list and malignant hyperthermia (MH) precautions were observed. Intraoperatively routine monitoring was used, warmed maintenance fluids were commenced, and a forced-air warming system was utilized to maintain temperature. The axillary block was performed with a 22G Stimuplex needle under ultrasound guidance with 20ml of 0.5% bupivacaine. After ensuring adequate anaesthesia of the limb, the tourniquet was inflated. Intravenous lignocaine 1% 3ml was given to minimize the pain of propofol infusion and a target-controlled infusion (TCI) of propofol using the Marsh Model to target a plasma concentration of 1-2 mcg.ml⁻¹ was commenced and titrated throughout the surgery aiming to achieve arousal to verbal stimulus. Surgery was uneventful with the tourniquet released at 59 minutes. The patient reported being comfortable in recovery and was safely discharged home the same day after routine day case discharge criteria were met. The patient was telephonically reviewed at days 1

and 2 postoperatively by the consultant anaesthetist. The block had fully resolved, pain was well controlled on oral analgesia and no post operative complications were noted.

Discussion

MC is a rare channelopathy caused by a mutation in *CLCN1* causing interference with the appropriate relaxation of skeletal muscle fibers after depolarization, resulting in prolonged muscle contraction or myotonia. The disease was first described by Thomas Thomsen in Schleswig, Denmark in 1876 who himself was affected by this disorder. Two main forms of MC are described: Thomsen's myotonia (AD) and Becker's myotonia (autosomal recessive) (2). Thomsen's myotonia presents with less severe symptoms of muscle stiffness predominantly affecting the upper limb muscles beginning from infancy to 2-3 years of age. Patients generally do not develop muscle weakness later in life. In contrast, Becker's myotonia is twice as common and presents later with more severe symptoms predominantly affecting the lower limbs. Symptoms are apparent between 4-12 years of age and can lead to muscle weakness later in life. Myotonia is often reported by patients as muscle stiffness, most pronounced in the extremities which ameliorates with continuous activity also known as the "warm-up phenomenon" and worsens after rest (3). The binding of acetylcholine to postsynaptic acetylcholine receptors causes the opening of sodium channels and sodium influx, leading to depolarization. The action potentials are propagated along the sarcolemma and t-tubule system. The arriving action potential, in turn, activates voltage-dependent calcium channels in the adjacent sarcoplasmic reticulum, causing a rise in cytosolic calcium levels and thus muscle contraction. Normally chloride influx stabilizes the action potential but in MC, this chloride conductance is impaired leading to prolonged contraction (4). Myotonic reactions in patients with MC may range from subclinical findings and periodic symptoms to disabling muscle weakness. Muscle stiffness may respond to sodium channel blockers such as mexiletine (currently the medication with the best documented effect), lamotrigine carbamazepine or phenytoin. Beneficial effects have also been reported with quinine, dantrolene, and acetazolamide (3).

In general, available literature outlining the anaesthetic considerations for the myotonic patient is relatively scarce. The few published case reports outlining anaesthetic considerations for these patients highlight potential issues of masseter spasm at induction leading to difficulty intubating, aspiration events and precipitation of myotonic contractions (4-7). Given these risks, MC is a condition that traditionally would have been considered a relative contraindication for day-case anaesthesia. Our case describes how a RA technique in combination with sedation using short acting drugs such as propofol, carefully titrated using a TCI can be used without any respiratory side effects and with careful planning and patient selection, the patient can be safely discharged home the same day of surgery.

Neither RA nor muscle relaxation can control myotonic contractions once they begin, emphasizing the importance of prevention. The anaesthetic plan for patients with MC must involve a consideration for minimizing triggers of myotonic reactions, some of which include hypothermia-induced shivering, pain (including pain on propofol injection), and certain drugs (4). Regional nerve block used in our report had the advantage of providing postoperative analgesia which is a key consideration for preventing myotonias in the postoperative period. Whilst a regional technique was used successfully here, the perioperative plan must include a plan for conversion to GA in the instance of a failed regional block. An awareness of the effects of the different neuromuscular blocking drugs on this condition is essential for the anaesthetist. Patients with MC are at high risk of developing

a severe myotonic response with generalized muscle spasms when depolarizing muscle relaxants (suxamethonium) are used. This could include masseter spasm, opisthotonos and decerebrate posturing making intubation and even ventilation difficult (7,8). In general, the response to non-depolarizing muscle relaxants (NDMRs) is reported to be normal, but dose reduction is advised in cases of established muscle weakness and wasting (late stages of Becker's disease). Anticholinesterase drugs have been shown to precipitate myotonia so short-acting NDMRs should be used when possible. Sugammadex for the reversal of rocuronium-induced neuromuscular block has been successfully employed but strong evidence suggesting its use is lacking. In vitro studies have shown that the administration of propofol had an anti-myotonic effect on skeletal muscle bundles but the clinical importance of this is unclear (9). Traditionally, concerns about susceptibility to MH have been described in patients with MC. Whilst this predisposition to MH is not clearly established, if sustained muscle contraction occurs during the induction or the course of anaesthesia, MH should always be suspected. Familiarity with MH protocol and availability of dantrolene should be ensured for all patients with MC presenting for anaesthesia.

Perioperative management of a patient with MC can be particularly challenging. Whilst no definitive recommendations exist recommending regional or GA for these patients, the combination of a regional anaesthetic technique and TCI of a short acting sedative agent should be considered.

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