Airway management and quadratus lumborum block for inguinal hernia repair in context of Chromosome 4q deletion: A case report and literature review

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Abstract

Chromosome 4q Deletion Syndrome (4QDS) is a rare chromosomal deletion that presents with various congenital defects that could affect anesthetic management. We present a patient with known 4QDS who underwent successful general anesthetic induction, tracheal intubation, and subsequently, bilateral quadratus lumborum nerve blocks, during laparoscopic inguinal hernia repair surgery.

Airway management and quadratus lumborum block for inguinal hernia repair in context of Chromosome 4q deletion: A case report and literature review

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Abstract: Chromosome 4q Deletion Syndrome (4QDS) is a rare chromosomal deletion with the potential for congenital defects that could affect the anesthetic management of affected patients. We present a report of a patient with known 4QDS who underwent successful general anesthetic induction and tracheal intubation, followed by subsequent bilateral quadratus lumborum nerve blocks, in the context of laparoscopic inguinal hernia repair surgery. Given the inconsistent presentation of patients with 4QDS reported in the literature, steps were taken to exercise sufficient caution concerning the overall management of this patient per the most recent guidelines for pediatric patients.

Key Clinical Message: Chromosome 4q Deletion Syndrome (4QDS) is a rare chromosomal deletion that presents with various congenital defects that could affect the anesthetic management. We present a patient with known 4QDS who underwent successful general anesthetic induction, tracheal intubation, and subsequently, bilateral quadratus lumborum nerve blocks, during laparoscopic inguinal hernia repair surgery.

Keywords : Chromosome 4q Deletion Syndrome, Congenital Airway Malformations, Challenging Pediatric Airway, Congenital Airway Malformations, Quadratus Lumborum Block, Regional Anesthesia

Introduction:

Deletion of the 4q chromosome is a rare chromosomal deletion syndrome estimated to occur in about 1 out of every 100,000 births.[1] The effects of this deletion present with various clinical phenotypes[2], such as congenital heart defects, craniofacial dysmorphic features, and developmental delay[1]. The gravity of physical characteristics is dependent on the amount and the site of chromatin that is missing. Most patients pass away before two years of age or, if they survive, are confronted with both social and physical barriers.[3]With specific reference to anesthesia, patients with this syndrome have inconsistently been reported to have laryngeal narrowing, raising the concern for intubation difficulties.[4,5] Due to the rarity of this syndrome and the myriad of clinical presentations, few guidelines are found for the induction of anesthesia in a patient with this syndrome.

In this report, we describe the anesthetic management of a patient with known chromosome abnormality of 46 XY, del(4)q34.1 - q35.2 and associated dysmorphic appearance, including micrognathia. Craniofacial abnormalities are of consideration in the selection of tracheal tube sizes and may result in difficult intubation. These same abnormalities can be severe, to the point in which patients require long-term tracheostomy or plastic/orthopedic surgery for jaw reconstruction.[6] In this report, we discuss the management of anesthesia, utilization of a quadratus lumborum plexus block, and provide images of the characteristic craniofacial abnormalities observed in Chromosome 4q Deletion Syndrome (4QDS). To our knowledge, no case to date has reported on the anesthetic management of pediatric patients with 4QDS, including nerve block procedures. Additionally, few resources in the literature exist that show clear images of the characteristic appearance of patients with 4QDS or document their intubation.

Case Report:

A 13-month-old male, 10.8 kg, presented for anesthesia pre-procedure assessment before a scheduled laparoscopic left inguinal hernia repair under general anesthesia. The patient was born at 36 weeks via spontaneous vaginal delivery. Echocardiogram at 30 hours of life revealed a patent foramen ovale (PFO) and patent ductus arteriosus (PDA) with associated left to right shunting. Outpatient follow-up with pediatric cardiology showed likely spontaneous defect closure and continued follow-up was not recommended. Past medical history was otherwise significant for genetic testing-confirmed chromosome 4q deletion (q34.1-35.2), constipation, arachnoid cysts, and bilateral hearing loss. This patient had no history of previous general anesthesia but had an uneventful sedation with propofol for MRI at an outside institution. At that time, the patient had a size 4 nasopharyngeal airway successfully placed in one attempt with no complications.

On physical exam, the patient had a moderately dysmorphic appearance, with slightly slanted palpebral fissures, flattened nasal root, low-set ears, left eyelid ptosis, high-arched palate with a triangular mouth and micrognathia (see Figure 1A and B). The patient's cardiovascular exam was unremarkable. The left inguinal hernia defect was easily reducible, and no signs of muscle weakness or lethargy were appreciated.

All other vital signs and electrocardiogram were unremarkable, and their airway was evaluated as not difficult. However, given the patient's history of 4QDS and micrognathia on examination, consideration was given for the patient to potentially require a smaller-than-expected tracheal tube, and thus, video laryngoscopy via C-MAC® was chosen for intubation. The patient's mother was informed of this plan and written consent was obtained. Shortly after the preoperative assessment, the patient was given 0.5 mg/kg of midazolam and 15 mg/kg of acetaminophen and then was brought to the operating room for general anesthesia induction, which was completed with propofol dosed at 3.2 mg/kg delivered in a 10 mg/mL bolus. Tracheal intubation was accomplished in one attempt via C-MAC® using a size 1 Miller blade and 3.5 ETT with microcuff. An intubating stylet and anterior, back, upward and right lateral pressure (BURP) were utilized to aid intubation. The laryngeal view was deemed Cormack-Lehane Grade I, with full visualization of the glottis. The view seen with the C-MAC® can be seen in Figure 2C and D, before and after intubation.

Laparoscopic repair of the patient's hernia commenced and was completed uneventfully. Upon closing the patient's surgical sites, two mL of 0.25% bupivacaine were injected into the umbilical incision. The anesthesia team then executed bilateral quadratus lumborum (QL) nerve blocks via ultrasound guidance. Needle placement was confirmed with saline injection and a total of 5 mg/kg of epinephrine and 0.8 mg/kg of ropivacaine were injected. After the QL nerve block, the patient was successfully extubated and transferred to the post-anesthesia care unit. Following surgery, the patient did not receive any additional medications for pain control and recovered uneventfully. The patient was discharged later the same day, and both three-day and one-month follow-up appointments were unremarkable; no complaints of pain were noted from the patient's mother beyond mild soreness on Day 3.

Discussion:

The deletion of the 4q chromosome is a rare deletion syndrome with only around 200 cases reported in the literature.[2] Based on the genetic analysis completed by Xu et al., the congenital heart defects associated with this syndrome have been localized to the 4q32.2-q34.3 region.[2] Our patient demonstrated deletions of the terminal end of this region (q34.1 - q35.2), which may have precipitated their PFO and PDA. The heart defects were visualized via echocardiogram on Day Two of life were evaluated and thought to likely close spontaneously without intervention. Upon preoperative physical exam by the anesthesia team, the patient's cardiovascular exam was grossly normal. However, close monitoring of their cardiovascular status was implemented due to their history of 4QDS. The patient's status was well-maintained throughout the procedure.

Our patient also presented with dysmorphic features associated with 4QDS such as flattening of the midface and nasal root, upturned nostrils, a high-arched palate, triangular mouth and posteriorly rotated, low-set ears - all features that could be concerning for intubation. Despite these dysmorphic features, the patient's ASA score was deemed a II and without concern for a difficult airway, which would suggest the patient was a good candidate for minimal, moderate or deep sedation[4]. However, considering the patient's anomalies, there was a concern for the airway requiring a smaller than expected ETT, especially since prior reports of 4QDS have demonstrated such.[4,5]

Per the 2022 American Society of Anesthesiologists (ASA), pediatric patients perceived as difficult or potentially difficult can be managed using an airway management algorithm with an emphasis on team-based care.[6] This care includes the use of video laryngoscopy to maximize visualization of a patient's airway and minimize airway intubation attempts. As such, video laryngoscopy via C-MAC® was chosen for intubation with success in one attempt. Dearlove and Sharples present a case report that highlighted a 4QDS patient whose laryngoscopic imaging demonstrated a small larynx with only the posterior arytenoids visualized.[4] While our patient presented with normal laryngeal anatomy as seen in Figure 1C and D, this report demonstrates the utility of video laryngoscopy for intubation in patients with 4QDS. There was potential for difficulty intubating our patient, and the video laryngoscope allowed the anesthesiologist to assess this in real time.

In addition to airway management, the anesthesia team also administered a QL block in the setting of laparoscopic inguinal hernia repair as a component of a multimodal anesthetic technique for pain management. Advances in pediatric regional anesthesia have evolved as the prevalence of ambulatory pediatric surgery has also grown. With the association of opioid use and perioperative complications, regional anesthesia has proven to be a safe and effective form of pain control in the ambulatory pediatric surgery setting, while also minimizing the use of narcotics.[7] When used in conjunction with Tylenol, as in our patient, this constitutes multimodal anesthetic technique, an approach with proven benefit in ambulatory pediatric patients.[8] While there is a documented report of pain insensitivity in 4QDS[9], our patient had previously demonstrated a response to painful stimuli in the setting of incarcerated inguinal hernia, which warranted the adoption of the regional anesthetic technique. The QL nerve block has demonstrated efficacy in multiple randomized control trials, presented by Zhao et al. in a meta-analysis of 346 patients, especially when administered under general anesthesia and ultrasound guidance.[10] The uncomplicated nature of our patient's nerve block, in addition to the lack of laryngeal narrowing seen during video-assisted intubation and the lack of persis-

tent cardiac defects, suggest the variance in presentation/phenotype present in patients with 4QDS. This is also in line with the inconsistent reporting of both pain insensitivity and laryngeal narrowing reported in the literature.[2,4,5,9]Our approach, with emphasis on the ASA 2022 Guidelines, maximized patient safety by emphasizing teamwork, communication, and caution. Some limitations of this approach, though, include the resources and time required to successfully carry out this plan. Smaller institutions may not have access to pediatric video laryngoscopy or providers trained in the use of such equipment.

Conclusion:

This case demonstrates successful intubation in a pediatric patient with 4QDS despite dysmorphic features and the potential for both laryngeal narrowing and pain insensitivity associated with the patient's genetic condition. While this patient was not expected to be a difficult airway upon preoperative physical examination, the dysmorphic facial features were a cause of concern and extra care was taken in patient intubation. In addition, a bilateral QL nerve block and adjunctive Tylenol, constituting multimodal anesthesia technique, were utilized in our patient for postoperative pain control. As demonstrated by this case, successful intubation and the use of both general and regional anesthesia can be safely accomplished in patients with 4QDS, though sufficient caution should be taken when assessing and treating this patient population. We suggest other patients with 4QDS be treated with similar caution, given the potential for adverse events related to inconsistently reported characteristics of this genetic syndrome.

Statement of Author Contribution:

Katie Lovell conducted chart review, figure creation, and completed the final report. **Michael C. Larkins** managed patient consent documentation, conducted chart review, and assisted with figure creation. **Melisa Pasli** assisted substantially with manuscript writing. **Adrienne H. Singleton** conceived the report, obtained patient consent and photography, and provided revisions and technical analysis of the report.

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No conflict of interest is declared by the authors.

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The authors declare that this manuscript is not published or under consideration in any other journals.

Consent:

Published with the written consent of the patient's mother.

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Figure Caption

Figure 1 - A: Frontal view of our patient with Chromosome 4q Deletion Syndrome; note the characteristic slightly slanted palpebral fissures, flattened nasal root, low-set ears and triangular mouth. B: Side profile of our patient, demonstrating the micrognathia associated with this syndrome. C: View of the patient's larynx via C-MAC® before tracheal intubation. D: The same view of our patient's larynx after intubation. No significant laryngeal abnormalities were noted during intubation.