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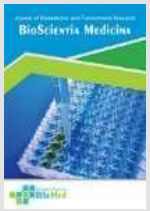

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Anesthetic Management of a Pediatric Patient with Popliteal Pterygium Syndrome Undergoing Labiopalatoplasty: A Case Report

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A B S T R A C T

Background: Popliteal pterygium syndrome (PPS) is a rare congenital disorder characterized by multiple anomalies, including orofacial, musculoskeletal, and genitourinary defects. Airway management in PPS patients can be challenging due to associated craniofacial abnormalities. This case report describes the anesthetic management of a 5-month-old infant with PPS undergoing labiopalatoplasty. **Case presentation:** A 5-month-old male infant, weighing 5.9 kg, presented for labiopalatoplasty. He had a diagnosis of PPS with associated labiopalatoschisis, ankyloblepharon filiforme, pterygium in the popliteal fossa, cryptorchidism, and syndactyly. Airway assessment revealed a patent airway with a cleft lip and palate. Anesthesia was induced with sevoflurane and maintained with sevoflurane and nitrous oxide. Direct laryngoscopy was unsuccessful, and videolaryngoscopy was used to facilitate tracheal intubation. **Conclusion:** This case highlights the challenges of airway management in infants with PPS. A thorough preoperative airway assessment and the availability of alternative intubation techniques, such as videolaryngoscopy, are crucial for successful anesthetic management in these patients.

1. Introduction

Popliteal pterygium syndrome (PPS), also known as facio-genito-popliteal syndrome, is a rare congenital disorder characterized by a constellation of anomalies primarily affecting the face, limbs, and genitalia. This syndrome, first described by Trelat in 1869, is caused by mutations in the interferon regulatory factor 6 (IRF6) gene, a critical regulator of ectodermal development. The IRF6 gene provides instructions for making a protein that plays a crucial role in the formation of skin, mucous membranes, and other tissues in the body. Mutations in this gene disrupt the normal development of these tissues, leading to the characteristic features of PPS. PPS is inherited in an autosomal dominant pattern, meaning that only one copy of the mutated gene is sufficient to cause the disorder. This pattern of inheritance suggests that if one parent has PPS, there is a 50% chance that each of their children will inherit the condition. However, due to the variability in the expression of the syndrome, some individuals may inherit the mutated gene but show only mild symptoms or even appear unaffected. The clinical presentation of PPS is highly variable, ranging from mild to severe. The hallmark features of the syndrome include popliteal pterygium (webbing behind the knees), cleft lip and/or palate, and genital abnormalities. Other associated anomalies may include syndactyly (fusion of fingers or toes),

contractures of various joints, lower lip pits or sinuses, and abnormalities of the eyelids, such as ankyloblepharon filiforme adnatum (fusion of the eyelids).[1-4]

The diagnosis of PPS is primarily based on clinical findings. The presence of two major criteria or one major and two minor criteria is usually sufficient to establish the diagnosis. Genetic testing can confirm the diagnosis and identify the specific mutation in the IRF6 gene. The management of PPS requires a multidisciplinary approach involving specialists from various fields, including pediatrics, genetics, plastic surgery, orthopedics, and ophthalmology. The treatment is tailored to the individual needs of the patient and may involve surgical correction of the various anomalies, physical therapy to improve joint mobility, and speech therapy to address speech difficulties associated with cleft lip and/or palate. Anesthesia for patients with PPS presents unique challenges due to the potential for difficult airway management. The craniofacial abnormalities, such as cleft lip and/or palate and micrognathia (small jaw), can make it difficult to visualize the larynx and intubate the trachea. In addition, the presence of syndactyly and contractures may limit access to the airway and make positioning the patient for surgery challenging.[5-7]

A thorough preoperative airway assessment is crucial for patients with PPS undergoing anesthesia. This assessment should include a detailed medical history, physical examination, and review of imaging studies. The anesthesiologist should be prepared for potential difficulties with mask ventilation and tracheal intubation. The availability of alternative intubation techniques, such as videolaryngoscopy and fiberoptic bronchoscopy, is essential.[8-10] In this case report, we describe the anesthetic management of a 5-month-old infant with PPS undergoing labiopalatoplasty.

2. Case Presentation

This case report details the anesthetic management of a 5-month-old male infant diagnosed with popliteal pterygium syndrome (PPS) who presented for labiopalatoplasty. The timeline of the disease, encompassing anamnesis, clinical findings, laboratory results, imaging findings, and the final diagnosis, is outlined below. The patient, a 5-month-old male infant, was referred to our hospital for corrective surgery for labiopalatoplasty. He had been previously diagnosed with PPS, a rare genetic disorder characterized by a variety of congenital anomalies. In this case, the infant presented with several key features associated with PPS; Labiopalatoschisis: A cleft lip and palate, a common craniofacial abnormality in PPS patients, was immediately apparent; Ankyloblepharon filiforme: This condition, characterized by fine fibrous bands connecting the upper and lower eyelids, was also observed; Popliteal Pterygium: The namesake feature of PPS, this involved the presence of webbed skin behind the knees, restricting movement and potentially impacting limb development; Cryptorchidism: The infant's medical history indicated undescended testicles, another anomaly frequently associated with PPS; Syndactyly: Fusion of the fingers or toes, a common musculoskeletal abnormality in PPS, was also noted in this patient. The infant's birth history was significant for a full-term delivery via cesarean section. Importantly, there was no history of cyanosis (bluish discoloration of the skin due to insufficient oxygen) or respiratory distress, suggesting adequate respiratory function despite the craniofacial anomalies. A comprehensive physical examination was conducted to assess the infant's overall health and identify any potential challenges for anesthesia and surgery. The key findings were; Airway: Despite the presence of a cleft lip and palate, the infant's airway was assessed as patent, meaning there was no obstruction to airflow. This is a critical observation for anesthetic planning, as a compromised airway can pose significant challenges during surgery; Breathing: Auscultation of the lungs revealed normal vesicular breath sounds, indicating clear and unobstructed airways and proper lung function. No wheezing or rhonchi (abnormal breath sounds) were detected,

further supporting healthy respiratory status; Circulation: The infant's cardiovascular system appeared normal, with regular heart sounds and no indications of any underlying cardiac abnormalities; Other: The physical examination confirmed the previously diagnosed features of PPS, including ankyloblepharon filiforme, popliteal pterygium, cryptorchidism, and syndactyly. These findings underscore the multi-systemic nature of PPS and the need for a comprehensive approach to patient care. A series of laboratory investigations were performed to assess the infant's overall health and identify any potential issues that might affect the surgical and anesthetic plan. The results were reassuringly normal; Complete blood count: Normal values indicated no evidence of infection, anemia, or other blood disorders; Electrolytes: Normal electrolyte levels are crucial for maintaining proper fluid balance and nerve and muscle function, essential for a safe perioperative course; Coagulation profile: Normal coagulation parameters ensured that the infant's blood clotting mechanisms were functioning correctly, minimizing the risk of excessive bleeding during and after surgery; Blood glucose: A normal blood glucose level indicated proper metabolic function and helped rule out any underlying endocrine disorders; Renal function tests: Normal renal function is important for eliminating waste products and medications from the body, contributing to a smooth recovery. Imaging studies provided a more detailed view of the infant's anatomy and helped identify any potential anatomical variations that might impact the surgical and anesthetic approach; Chest X-ray: An unremarkable chest X-ray confirmed normal lung structure and ruled out any underlying respiratory conditions; Craniofacial CT scan: This scan revealed a narrowing of the nasopharynx and oropharynx, the upper parts of the airway behind the nose and mouth. This finding is relevant for airway management, as it could potentially complicate mask ventilation or tracheal intubation; Echocardiogram: This test, which uses sound waves to create images of the heart, showed no congenital heart defects. This is an important consideration for anesthesia, as underlying cardiac issues can increase the risks associated with surgery; Skeletal survey: This comprehensive X-ray examination of the bones revealed mild scoliosis (curvature of the spine) and delayed bone age. While these findings may not directly impact the immediate anesthetic management, they provide valuable information for long-term monitoring and management of the patient's musculoskeletal development. Based on the constellation of clinical features, supported by the laboratory and imaging findings, the diagnosis of popliteal pterygium syndrome (PPS) with associated labiopalatoschisis was confirmed. This diagnosis informed the development of a comprehensive anesthetic and surgical plan tailored to the specific needs and challenges presented by this rare condition (Table 1).

The primary surgical intervention for this patient was a labiopalatoplasty, a procedure aimed at correcting the cleft lip and palate. This is a complex surgical procedure that requires meticulous technique and careful attention to detail; Labiopalatoplasty: The surgery was performed by a qualified and experienced pediatric plastic surgeon specializing in craniofacial procedures. The operation was conducted under general anesthesia, as described in the previous section, and lasted for approximately 90 minutes. Throughout the procedure, the surgical team maintained a focus on minimizing tissue trauma and optimizing aesthetic outcomes. Importantly, there were no intraoperative complications reported, indicating a smooth surgical course. Following the completion of the labiopalatoplasty, the infant was transferred to the Post-Anesthesia Care Unit (PACU) for close observation and monitoring during the immediate postoperative period; PACU Observation: In the PACU, the infant's vital signs were closely monitored, including heart rate, blood pressure, oxygen saturation, and respiratory rate. This continuous monitoring allowed the medical team to promptly detect and address any potential complications, such as respiratory distress, bleeding, or hemodynamic instability. Pain management was

also prioritized, with analgesics administered as needed to ensure the infant's comfort and facilitate recovery. The PACU team remained vigilant in assessing for any signs of complications, such as bleeding, infection, or airway obstruction, which can occur following labiopalatoplasty; Discharge and Ward Management: Once the infant was deemed stable, as indicated by stable vital signs, adequate pain control, and no signs of complications, he was discharged to the pediatric ward for continued care. A Steward score of 5, indicating a high level of recovery and readiness for discharge, was documented. On the ward, nursing staff provided ongoing monitoring and support, including pain management, wound care, and feeding assistance; Long-term Follow-up: To ensure optimal healing and address any potential long-term complications, further follow-up appointments were scheduled with the surgical team. These appointments allowed for ongoing assessment of the surgical site, monitoring for any signs of infection or dehiscence (wound separation), and evaluation of the infant's feeding and speech development. Long-term follow-up is crucial in patients with PPS, as they may require ongoing multidisciplinary care to address the various manifestations of the syndrome. This detailed description of the surgical procedure and postoperative follow-up highlights the comprehensive care provided to this infant with PPS. The meticulous attention to detail and the multidisciplinary approach are essential for optimizing outcomes in these complex cases (Table 2).

Given the complex nature of PPS and the potential for airway difficulties, meticulous preoperative planning was essential. The anesthetic team undertook a comprehensive assessment and preparation process; Evaluation of Medical History and Airway: A thorough review of the infant's medical records, including the previously discussed clinical and imaging findings, was conducted. Particular attention was paid to the airway examination, noting the presence of the cleft lip and palate and the potential for narrowing of the nasopharynx and oropharynx as indicated by the CT scan. This information guided the formulation of an individualized anesthetic plan; Discussion of Risks and Benefits: The anesthetic team engaged in a detailed discussion with the infant's parents, explaining the potential risks and benefits associated with general anesthesia in the context of PPS and labiopalatoplasty. This informed consent process ensured that the parents were fully aware of the potential challenges and could actively participate in the decision-making process; Planning for Potential Airway Difficulties: Anticipating potential challenges with airway management, the team prepared for various scenarios. This included having a range of airway equipment readily available, such as different sizes of endotracheal tubes (ETTs), laryngoscope blades (Macintosh and Miller), and a videolaryngoscope (GlideScope) to facilitate intubation if direct laryngoscopy proved difficult; Premedication: To facilitate a smooth induction of anesthesia and minimize anxiety, the infant received premedication with ketamine (1 mg/kg IV) and atropine (0.1 mg/kg IV). Ketamine provides sedation and analgesia, while atropine helps to reduce secretions and prevent bradycardia (slow heart rate) during anesthesia. The anesthetic management focused on ensuring a safe and stable anesthetic course while addressing the potential airway challenges associated with PPS; Induction: Anesthesia was induced via inhalation with sevoflurane in a mixture of oxygen and nitrous oxide. This technique allows for a gradual and controlled induction, minimizing the risk of airway complications. Once an adequate depth of anesthesia was achieved, positive pressure ventilation was initiated to ensure adequate oxygenation and ventilation. Intravenous fentanyl (2 mcg/kg) was administered for analgesia, and atracurium (0.5 mg/kg) was given to provide muscle relaxation, facilitating tracheal intubation; Airway Management: As anticipated, direct laryngoscopy using both Macintosh and Miller blades was unsuccessful in visualizing the glottis due to the anatomical challenges posed by the cleft palate and potentially narrowed upper airway. Recognizing the difficulty, the team

promptly switched to the GlideScope videolaryngoscope, which provided an improved view of the glottis, enabling successful tracheal intubation. An uncuffed ETT size of 3.0 mm was chosen, considering the infant's age and the potential for tracheal narrowing in PPS. The ETT was secured at a depth of 9 cm, and its correct placement was confirmed by auscultation of bilateral breath sounds and capnography ($ETCO_2$) tracing; Maintenance: Anesthesia was maintained with sevoflurane and nitrous oxide in oxygen, titrated to maintain an appropriate depth of anesthesia. Throughout the procedure, the infant's vital signs, including heart rate, blood pressure, oxygen saturation, and $ETCO_2$, were continuously monitored to ensure hemodynamic stability and adequate ventilation. The concluding phase of anesthetic management focused on a smooth emergence from anesthesia and a safe transition to the postoperative period; Reversal of Neuromuscular Blockade: At the end of the surgical procedure, the neuromuscular blockade induced by atracurium was reversed with an appropriate dose of neostigmine and glycopyrrolate. This ensured the return of adequate muscle strength and spontaneous respiration before extubation; Extubation: Once the infant regained sufficient consciousness and spontaneous ventilation was established, the ETT was removed in the operating room. The infant was closely observed for any signs of respiratory distress or airway obstruction following extubation; PACU Observation: The infant was then transferred to the PACU for continued monitoring and observation. The PACU team assessed the infant's vital signs, pain level, and level of consciousness, ensuring a smooth recovery from anesthesia. A Steward score of 5, indicating a high level of recovery, was documented before transfer to the pediatric ward; Transfer to Pediatric Ward: Once stable, the infant was transferred to the pediatric ward for ongoing care. The ward team continued to monitor the infant's recovery, providing pain management, wound care, and feeding support. This detailed account of anesthetic management demonstrates the importance of a comprehensive and adaptable approach in patients with PPS. The successful management of this case highlights the value of thorough preoperative planning, the availability of alternative airway management techniques, and close monitoring throughout the perioperative period (Table 3).

Table 1. Timeline of the disease, including anamnesis, clinical findings, laboratory and imaging findings, and the diagnosis.

Category	Findings
Anamnesis	- 5-month-old boy. - Referred for labiopalatoplasty. - Diagnosed with popliteal pterygium syndrome (PPS). - Associated features: labiopalatoschisis, ankyloblepharon filiforme, pterygium in the popliteal fossa, cryptorchidism, and syndactyly. - Born at term via cesarean delivery. - No history of cyanosis or respiratory distress.
Clinical finding	- Airway: Patent airway with cleft lip and palate. - Breathing: Vesicular breath sounds, no wheezing or rhonchi. - Circulation: Normal heart sounds. - Other: Ankyloblepharon filiforme, pterygium in the popliteal fossa, cryptorchidism, and syndactyly.
Imaging	- Chest X-ray: Unremarkable - Craniofacial CT scan: Narrowing of the nasopharynx and oropharynx. - Echocardiogram: No congenital heart defects detected. - Skeletal survey: Mild scoliosis, delayed bone age.
Laboratory	- Complete blood count: Normal - Electrolytes: Normal - Coagulation profile: Normal - Blood glucose: 80 mg/dL - Renal function tests: Within normal limits
Diagnosis	- Popliteal pterygium syndrome (PPS) - Labiopalatoschisis

Table 2. Procedure of surgery and follow-up after surgery procedure.

Category	Details
Surgical procedure	Labiopalatoplasty - The surgical procedure was performed by a qualified surgeon. - The procedure lasted for 90 minutes. - There were no intraoperative complications reported.
Postoperative follow-up	- The patient was transferred to the PACU for observation. - Vital signs were monitored, including heart rate, blood pressure, oxygen saturation, and respiratory rate. - Pain management was provided as needed. - The patient was assessed for any signs of complications, such as bleeding, infection, or airway obstruction. - Once stable, the patient was discharged to the pediatric ward with a Steward score of 5. - Further follow-up appointments were scheduled to monitor healing and assess for any long-term complications.

Table 3. Preoperative management, anesthetic management, postoperative management.

Category	Details
Preoperative	- Evaluation of the patient's medical history and airway. - Discussion of potential risks and benefits of anesthesia. - Planning for potential airway difficulties. - Premedication with ketamine (1 mg/kg IV) and atropine (0.1 mg/kg IV).
Induction	- Inhalation induction with sevoflurane in oxygen and nitrous oxide. - Positive pressure ventilation. - Administration of fentanyl (2 mcg/kg IV) and atracurium (0.5 mg/kg IV).
airway management	- Direct laryngoscopy with Macintosh and Miller blades unsuccessful. - Videolaryngoscopy with GlideScope used for successful intubation. - Uncuffed ETT size 3.0 mm used. - ETT secured at a depth of 9 cm. - Confirmation of ETT placement by auscultation and $ETCO_2$.
maintenance	- Anesthesia maintained with sevoflurane and nitrous oxide in oxygen. - Monitoring of vital signs, including heart rate, blood pressure, oxygen saturation, and $ETCO_2$.
emergence and postoperative	- Reversal of neuromuscular blockade. - Extubation in the operating room. - Observation in the PACU. - Steward score of 5 in the PACU. - Transfer to the pediatric ward.

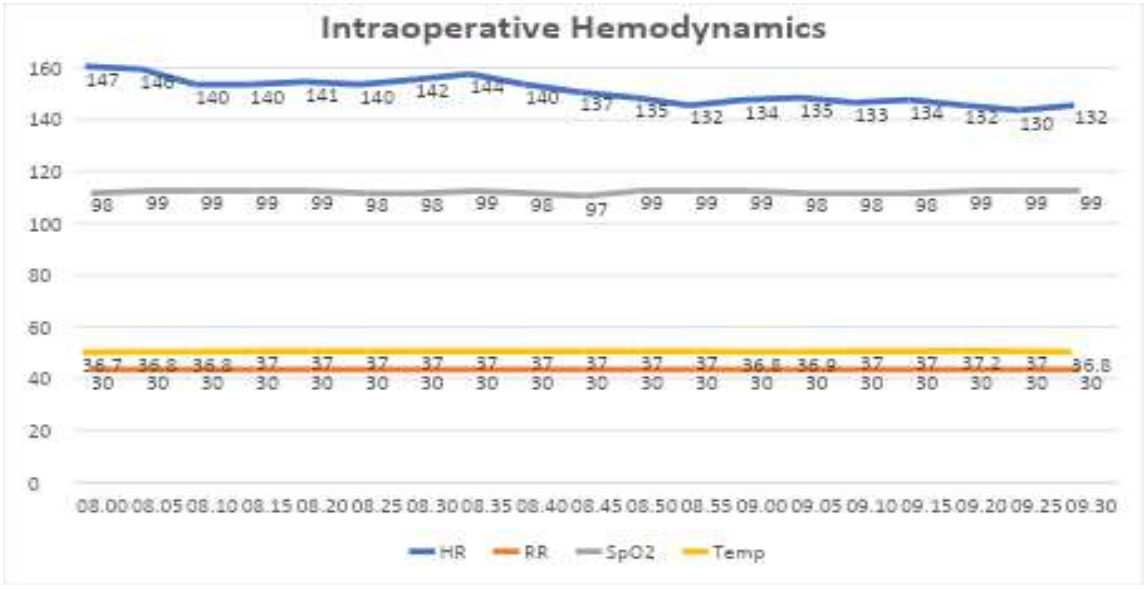

Figure 1. Intraoperative hemodynamics.

3. Discussion

Airway management in pediatric patients with popliteal pterygium syndrome (PPS) presents unique challenges for anesthesiologists. The syndrome's hallmark craniofacial anomalies can significantly complicate the process of establishing and maintaining a patent airway, making it a critical aspect of perioperative care for these patients. PPS is characterized by a spectrum of abnormalities, with micrognathia (small jaw), cleft palate, and glossoptosis (posterior displacement of the tongue) being particularly relevant to airway management. These anomalies can impede both bag-mask ventilation and endotracheal intubation, increasing the risk of airway obstruction and complications. Micrognathia, a condition characterized by a smaller-than-normal jaw, is a common craniofacial anomaly encountered in patients with popliteal pterygium syndrome (PPS). This seemingly subtle deviation in facial structure can have significant implications for airway management, particularly during procedures requiring general anesthesia and tracheal intubation. In the context of PPS, micrognathia presents a unique set of challenges for anesthesiologists. The reduced size of the mandible (lower jaw) alters the spatial relationship between key anatomical structures in the airway, making it more difficult to establish and maintain a patent airway. Direct laryngoscopy, a cornerstone technique in airway management, involves inserting a laryngoscope blade into the mouth to visualize the glottis and facilitate tracheal intubation. However, in patients with micrognathia, the reduced size of the jaw limits the space available for maneuvering the laryngoscope blade. This restriction can make it challenging to achieve an optimal view of the glottis, increasing the difficulty of guiding the endotracheal tube into the trachea. The small jaw size can impede the proper placement and manipulation of the laryngoscope blade. The blade may not sit correctly within the oral cavity, making it difficult to lift the tongue and epiglottis to expose the glottis. This can lead to multiple attempts at laryngoscopy, increasing the risk of trauma to the surrounding tissues and teeth. Even with proper placement of the laryngoscope blade, micrognathia can still hinder the visualization of the glottis. The altered anatomical configuration may obscure the view of the glottis, making it difficult to identify the landmarks necessary for successful intubation. This limited visualization can increase the risk of misplacement of the endotracheal tube, potentially leading to complications like esophageal intubation or airway obstruction. The challenges

associated with micrognathia in PPS can increase the risk of complications during tracheal intubation. Multiple attempts at laryngoscopy can lead to trauma to the airway, including mucosal tears, bleeding, and dental damage. Additionally, the difficulty in visualizing the glottis can increase the risk of hypoxia, a condition caused by insufficient oxygen supply to the body. Recognizing the potential difficulties associated with micrognathia, anesthesiologists often consider alternative airway management techniques in PPS patients. Videolaryngoscopy, a technique that uses a video camera to visualize the glottis, can be particularly helpful in these cases. The enhanced view provided by videolaryngoscopy can improve the success rate of intubation and reduce the risk of complications. Cleft palate, a congenital anomaly characterized by a gap or opening in the roof of the mouth, is another prevalent feature of popliteal pterygium syndrome (PPS) that can significantly complicate airway management. This disruption in the normal anatomy of the palate poses unique challenges for anesthesiologists, particularly during procedures requiring general anesthesia and tracheal intubation. In individuals with a cleft palate, the separation between the oral and nasal cavities disrupts the normal airflow pathway. This disruption can make it difficult to achieve an effective seal around the face mask during bag-mask ventilation, a crucial step in providing oxygen and anesthetic gases before and after intubation. The gap in the palate creates a leak that prevents a complete seal between the face mask and the patient's face. This leak can result in a significant loss of tidal volume, the amount of air that moves in and out of the lungs with each breath. As a consequence, the effectiveness of ventilation is reduced, making it challenging to maintain adequate oxygenation and achieve an appropriate depth of anesthesia. The inability to achieve a proper mask seal can lead to inadequate ventilation and oxygenation. The leak allows anesthetic gases and oxygen to escape into the atmosphere, reducing the amount delivered to the lungs. This can result in hypoxia (low oxygen levels) and hypercarbia (high carbon dioxide levels), potentially leading to complications during anesthesia. The leak in the mask seal can also make it difficult to maintain an appropriate depth of anesthesia. Anesthetic gases, such as sevoflurane or desflurane, may escape through the gap in the palate, reducing the concentration delivered to the lungs and making it challenging to keep the patient adequately anesthetized. Anesthesiologists employ various strategies to address the challenges posed by cleft palate during airway management. Choosing a mask that fits snugly around the patient's face and carefully positioning it to minimize the leak can improve the effectiveness of ventilation. Inserting an oral airway can help to maintain a patent airway and improve airflow during mask ventilation. In some cases, early tracheal intubation may be considered to secure the airway and ensure adequate ventilation and oxygenation. Supraglottic airway devices, such as the laryngeal mask airway (LMA), may be used as an alternative to face mask ventilation or tracheal intubation in certain situations. Glossoptosis, the posterior displacement or retraction of the tongue, is another significant airway challenge encountered in patients with popliteal pterygium syndrome (PPS). This anatomical abnormality, where the tongue base falls back towards the posterior wall of the pharynx, can lead to partial or complete obstruction of the airway, making it a critical consideration in anesthetic management. In individuals with PPS, glossoptosis can occur due to a combination of factors, including mandibular hypoplasia (underdevelopment of the lower jaw), hypotonia (reduced muscle tone) of the tongue muscles, and the presence of a cleft palate. These factors can contribute to the posterior displacement of the tongue, increasing the risk of airway obstruction. Glossoptosis can impede both spontaneous breathing and assisted ventilation. In spontaneous breathing, the retracted tongue can partially or completely block the flow of air through the pharynx, making it difficult for the patient to inhale and exhale effectively. This can lead to hypoxia (low oxygen levels) and hypercarbia (high carbon dioxide levels), potentially causing respiratory distress and

complications during anesthesia. Similarly, glossoptosis can also hinder assisted ventilation, such as bag-mask ventilation or mechanical ventilation. The retracted tongue can create resistance to airflow, making it challenging to deliver adequate tidal volumes and maintain proper oxygenation and ventilation. Anesthesiologists employ various strategies to manage glossoptosis in PPS patients and maintain a patent airway. Proper head positioning, such as extending the neck and tilting the head back, can help to bring the tongue forward and relieve the obstruction. The jaw thrust maneuver, which involves manually displacing the jaw forward, can also help to open the airway and improve airflow. The use of airway adjuncts, such as oral airways or nasal airways, can help to maintain a patent airway and prevent the tongue from falling back and obstructing the airflow. In some cases, early tracheal intubation may be considered to secure the airway and ensure adequate ventilation and oxygenation, especially if mask ventilation proves difficult. Continuous monitoring of oxygen saturation, respiratory rate, and end-tidal carbon dioxide levels is crucial in patients with glossoptosis. Anesthesiologists must remain vigilant and be prepared to intervene promptly if there are signs of airway obstruction or respiratory compromise. Syndactyly, a congenital anomaly characterized by the fusion of fingers or toes, is a musculoskeletal manifestation of PPS that can indirectly impact airway management. While it may not directly obstruct the airway, syndactyly can limit the dexterity and flexibility of the anesthesiologist's hands, making it more challenging to perform delicate procedures like laryngoscopy and intubation. The restricted finger movement caused by syndactyly can hinder the ability to manipulate the laryngoscope blade and endotracheal tube effectively. This can make it more difficult to achieve an optimal view of the glottis, guide the endotracheal tube into the trachea, and perform other airway management procedures. During laryngoscopy, the anesthesiologist needs to hold the laryngoscope blade with a firm and steady grip while carefully manipulating it to lift the tongue and epiglottis to expose the glottis. Syndactyly can make it challenging to maintain a secure grip on the laryngoscope blade and perform these delicate maneuvers effectively. Similarly, during intubation, the anesthesiologist needs to guide the endotracheal tube through the glottis and into the trachea. This requires precise hand movements and coordination. Syndactyly can limit the flexibility and range of motion of the fingers, making it more difficult to guide the endotracheal tube accurately. While syndactyly may not make airway management impossible, it can increase the difficulty of the procedure, potentially leading to multiple attempts, trauma to the airway, and complications like hypoxia. Anesthesiologists should be aware of this potential challenge and be prepared to adjust their technique or consider alternative airway management approaches if necessary. Contractures, another musculoskeletal manifestation of PPS, involve the tightening or shortening of muscles or tendons around joints, limiting their range of motion. These contractures can pose significant challenges for airway management by making it difficult to position the patient optimally for surgery and intubation. Proper patient positioning is crucial for successful airway management. For example, extending the neck and tilting the head back can help to align the airway axes and improve visualization of the glottis during laryngoscopy. However, contractures in the neck or jaw can restrict head extension, making it challenging to achieve optimal positioning for intubation. Contractures can also limit the ability to perform certain airway maneuvers, such as the jaw thrust maneuver, which is often used to open the airway and improve airflow. Contractures in the temporomandibular joint (TMJ) can restrict jaw movement, making it difficult to perform this maneuver effectively. The presence of contractures can increase the difficulty of airway management and potentially lead to complications. Multiple attempts at laryngoscopy or intubation may be required, increasing the risk of trauma to the airway. Additionally, the difficulty in achieving optimal positioning and performing airway maneuvers can

increase the risk of hypoxia and other complications.[11-14]

A thorough preoperative airway assessment is of paramount importance for all patients with popliteal pterygium syndrome (PPS) scheduled to undergo anesthesia. This comprehensive evaluation serves as the cornerstone for developing a safe and effective anesthetic plan, considering the unique airway challenges associated with this syndrome. PPS, characterized by a spectrum of craniofacial and musculoskeletal anomalies, can significantly complicate airway management, making a meticulous preoperative assessment crucial for anticipating and mitigating potential difficulties. The preoperative airway assessment in PPS patients should encompass a detailed review of the patient's medical history, a comprehensive physical examination, and appropriate imaging studies. Each component of this assessment provides valuable information that contributes to a thorough understanding of the patient's airway anatomy and potential challenges. The review of the patient's medical history is the first step in the preoperative airway assessment. This involves gathering relevant information about the patient's past medical and anesthetic experiences, with a particular focus on any history of difficult airway management. Accessing and reviewing any previous anesthetic records can offer invaluable insights into the patient's airway history. These records often contain detailed information about past anesthetic encounters, including any challenges faced during airway management. Pay close attention to documentation regarding previous intubation attempts, the techniques employed (direct laryngoscopy, videolaryngoscopy, etc.), the ease or difficulty of intubation, and any complications encountered (e.g., esophageal intubation, airway trauma, desaturation). This information allows the anesthesiologist to anticipate potential challenges in the current setting and proactively plan for alternative airway management strategies if needed. A documented history of difficult airway management, whether in the context of previous surgeries, emergency procedures, or critical care situations, serves as a crucial warning sign. This information should prompt a heightened awareness of the potential for similar difficulties in the current case and trigger a more detailed investigation of the airway. Understanding the nature of previous difficulties (e.g., difficult mask ventilation, difficult laryngoscopy, inability to intubate) can guide the selection of appropriate airway equipment and techniques. Certain underlying medical conditions can significantly influence airway management. These include respiratory disorders (e.g., asthma, bronchopulmonary dysplasia), cardiovascular diseases (e.g., congenital heart defects), neuromuscular disorders (e.g., muscular dystrophy), and genetic syndromes associated with airway abnormalities. It is essential to obtain a comprehensive medical history, including a review of all current and past medical conditions, to assess their potential impact on the anesthetic plan. For example, a patient with a history of severe asthma may require pre-operative bronchodilator therapy or a modified anesthetic technique to minimize the risk of bronchospasm. Some medications can have direct or indirect effects on airway management. These include medications that affect muscle tone (e.g., muscle relaxants), those with potential respiratory depressant effects (e.g., opioids, sedatives), and those that may interact with anesthetic agents. A thorough medication history, including prescription medications, over-the-counter drugs, and herbal supplements, is necessary to identify any potential drug interactions or adverse effects that may complicate anesthesia. For instance, if a patient is taking a muscle relaxant for a neuromuscular disorder, the anesthesiologist may need to adjust the dose of neuromuscular blocking agents used during anesthesia to avoid prolonged paralysis. The physical examination is a crucial component of the preoperative airway assessment, providing a direct evaluation of the patient's airway anatomy and identifying any specific features that may pose challenges during airway management. Assessing the

patient's mouth opening, typically measured as the interincisor distance (the distance between the upper and lower incisors), provides an indication of the space available for inserting a laryngoscope blade during direct laryngoscopy. A restricted mouth opening, often associated with micrognathia or TMJ (temporomandibular joint) limitations, can make it challenging to visualize the glottis and intubate the trachea. In such cases, alternative intubation techniques, such as videolaryngoscopy or fiberoptic intubation, may be necessary. The thyromental distance, measured from the tip of the thyroid cartilage (Adam's apple) to the tip of the chin with the neck fully extended, reflects the length of the mandibular space. A short thyromental distance can indicate a limited space for laryngeal manipulation and intubation, potentially making it difficult to achieve an optimal view of the glottis. The Mallampati score is a widely used classification system that helps predict the ease of direct laryngoscopy. It involves visualizing the oropharyngeal structures with the patient's mouth wide open and tongue protruding to the maximum extent. The score is based on the visibility of the soft palate, uvula, faucial pillars, and base of the tongue. A higher Mallampati score (Class III or IV) suggests a greater likelihood of difficult laryngoscopy. Assessing the patient's neck mobility, including flexion, extension, and lateral rotation, is essential for determining the ease of positioning for intubation. Limited neck mobility, often associated with cervical spine abnormalities or contractures, can hinder proper positioning and make it challenging to align the airway axes for successful intubation. The presence of other craniofacial anomalies, such as micrognathia (small jaw), retrognathia (receding jaw), macroglossia (enlarged tongue), or cleft palate, can further complicate airway management. These anomalies should be carefully documented and considered during the anesthetic planning, as they may require modifications in the intubation technique or the use of alternative airway devices. Imaging studies can provide valuable information about the patient's airway anatomy, helping to identify any anatomical abnormalities that may contribute to difficult airway management. A CT scan of the head and neck can provide detailed images of the bony and soft tissue structures of the airway, offering a comprehensive view of the airway anatomy. This can help identify any narrowing, deviation, or obstruction in the airway that may complicate intubation. For example, a CT scan can reveal tracheal stenosis, a condition where the trachea is abnormally narrowed, which may require the use of a smaller endotracheal tube. An MRI can provide additional information about the soft tissues of the airway, such as the tongue, epiglottis, and vocal cords. This can be helpful in assessing the degree of glossoptosis or other soft tissue abnormalities that may contribute to airway obstruction. MRI is particularly useful for evaluating the extent of soft tissue involvement in conditions like Pierre Robin sequence, which is characterized by micrognathia, glossoptosis, and cleft palate. A lateral neck X-ray can provide a basic assessment of the airway, including the size and shape of the trachea and the position of the epiglottis. This can be helpful in identifying any gross abnormalities that may warrant further investigation with more advanced imaging techniques. While a lateral neck X-ray may not be as detailed as a CT scan or MRI, it can be a useful initial screening tool for identifying potential airway concerns.[15-17]

Anesthetic management in patients with popliteal pterygium syndrome (PPS) requires a comprehensive and individualized approach, considering the unique challenges posed by this rare congenital disorder. PPS is characterized by a variety of abnormalities, including craniofacial anomalies, musculoskeletal anomalies, and other systemic involvement. These abnormalities can significantly impact airway management, making careful planning and preparation essential for ensuring a safe and successful anesthetic course. The anesthetic management of PPS patients should be tailored to the individual needs of each patient and the planned surgical procedure. Factors to consider include the patient's age, overall health status, the presence of

specific anomalies, and the nature and duration of the surgery. It is crucial to be prepared for potential difficulties with airway management in PPS patients. The presence of craniofacial anomalies, such as micrognathia (small jaw), cleft palate, and glossoptosis (posterior displacement of the tongue), can make it difficult to mask ventilate, intubate the trachea, and maintain a patent airway. A variety of airway management techniques may be used in PPS patients, depending on the specific airway challenges encountered. Direct laryngoscopy is the most common technique for tracheal intubation, involving the use of a laryngoscope blade to visualize the glottis and guide the endotracheal tube into the trachea. However, direct laryngoscopy may be difficult in PPS patients with craniofacial anomalies that limit visualization of the glottis. Videolaryngoscopy technique uses a video camera attached to a laryngoscope blade to provide an enhanced view of the glottis. Videolaryngoscopy can be helpful in patients with difficult airways, as it allows for better visualization of the glottis and can facilitate intubation. Fiberoptic bronchoscopy technique involves the use of a flexible fiberoptic scope to visualize the airway and intubate the trachea. Fiberoptic bronchoscopy can be used in patients with very difficult airways, such as those with severe micrognathia or glossoptosis. Supraglottic airway devices such as the laryngeal mask airway (LMA), can be used to provide ventilation and oxygenation in patients who cannot be intubated. Supraglottic airway devices are inserted into the pharynx and sit above the glottis, providing a seal for ventilation without the need for tracheal intubation. PPS patients should be closely monitored in the postoperative period for any signs of respiratory distress. This includes monitoring the patient's oxygen saturation, respiratory rate, and work of breathing. PPS patients may also be at increased risk for postoperative complications, such as bleeding and infection. It is important to monitor the patient closely for these complications. In addition to the challenges of airway management, PPS patients may also have other medical conditions that need to be considered during the perioperative period. PPS patients may have congenital heart defects, which can increase the risk of complications during anesthesia. PPS patients may have gastrointestinal abnormalities, such as esophageal atresia or tracheoesophageal fistula. These abnormalities can make it difficult to place a nasogastric tube or to feed the patient. PPS patients may have genitourinary abnormalities, such as hypospadias or cryptorchidism. These abnormalities may require surgical correction.[18-20]

4. Conclusion

This case report presents the successful anesthetic management of a 5-month-old infant with PPS undergoing labiopalatoplasty. It highlights the importance of a thorough preoperative airway assessment and preparedness for difficult airway management in such cases. The use of videolaryngoscopy proved crucial in successfully securing the airway, emphasizing the value of having alternative intubation techniques readily available. PPS patients often require multiple surgical procedures throughout their lives, necessitating careful anesthetic planning each time. This case underscores the need for anesthesiologists to be well-versed in the airway challenges associated with PPS and to remain adaptable in their approach to airway management.

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