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Case report

Respiratory failure after superior-based pharyngeal flap for velopharyngeal insufficiency: A rare complication☆

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ABSTRACT

Velopharyngeal insufficiency (VPI) is an uncommon pediatric disorder often associated with congenital syndromes. After speech therapy, surgery is the standard management. Many surgical approaches to VPI repair have been reported and the complications of these procedures are well documented. To date, there have been no published cases of respiratory failure secondary to pneumomediastinum, pneumopericardium, and bilateral pneumothoraces with associated subcutaneous emphysema after superior-based pharyngeal flap. We present the first case in the literature. Our proposed etiology for the respiratory failure is air tracking from the flap donor site to the pleural spaces of the thoracic cavity via the visceral or prevertebral fascia following positive pressure ventilation.

1. Introduction

Velopharyngeal insufficiency (VPI) is a disorder characterized by incomplete closure of the velopharyngeal sphincter during speech and swallowing [1]. The result is a patient with hypernasal speech, increased nasal resonance, and in some instances, nasal regurgitation of liquids and solids. Other common associations are poor speech intelligibility and speech delay [2]. The etiology can be congenital, associated with anatomic, functional, or neuromuscular abnormalities, or acquired during adenoidectomy [1,3]. The precise incidence of congenital VPI has not been reported in the literature but there is a known association between VPI and pediatric syndromes, of note velocardiofacial syndrome, Pierre-Robin, and cleft palate [1]. Thorough consideration of a syndromic child and their associated constellation of abnormalities is critical in the preoperative planning for VPI repair.

A multidisciplinary team including speech therapists, pediatric otolaryngologists, and prosthodontists participate in the evaluation and diagnosis of VPI [1,3]. Patients typically undergo a trial of speech therapy for 6 months to determine if they have the compensatory mechanisms necessary to pursue non-operative management. If a patient continues to have hypernasal speech, surgical repair is planned when the child is aged 3 or 4 years [1]. Numerous approaches to the surgical repair of VPI have been reported, including superior-based pharyngeal flap, sphincter pharyngoplasty, Furlow palatoplasty, and augmentation pharyngoplasty, and calcium hydroxylapatite or autologous fat injection [1–4]. Selection of which procedure(s) best suits each patient is highly individualized, taking into consideration suspected etiology, anatomic gap, age, syndromes, and many other factors.

Outcomes of VPI repair are measured by improvement in hypernasal speech and post-operative complications. Complications documented in the literature include bleeding, aspiration, airway obstruction requiring reintubation/tracheotomy, flap dehiscence, and nasopharyngeal stenosis; however, the complication that has received the most attention is post-operative obstructive sleep apnea [1,4]. To our knowledge, respiratory failure has only been documented twice in recent literature and it was related to a central nervous system insult [5]. We present here a unique etiology of respiratory failure following superior-based pharyngeal flap.

2. Case report

Institutional review board exemption was obtained from our institution. Our patient was a 3-year, 6-month-old female referred to the pediatric otolaryngology clinic for evaluation of speech delay, nasal speech, and possible velopharyngeal insufficiency. Her medical history was significant for an unremarkable gestation, born at 37 weeks weighing 4 lb 4 oz, and a 3 week neonatal

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intensive care unit (ICU) stay at birth for congestive heart failure related to patent ductus arteriosus (PDA) and ventriculoseptal defect (VSD). She underwent PDA coiling at 9 months of age. Her VSD closed spontaneously. She had no history of cleft lip/palate or any other craniofacial anomalies though her facial features were noted to be dysmorphic. Genetics work up was negative for the 22q11 deletion. Speech pathology evaluation revealed severe nasality and speech delay consistent with VPI. She underwent two years of speech therapy with little improvement prior to ENT evaluation. Our initial exam was significant for 3+ tonsils and moderate adenoid tissue. Nasopharyngoscopy demonstrated good coronal closure but persistent 2–3 cm central gap with attempted sagittal velopharyngeal closure. Our patient underwent tonsillectomy without adenoidectomy 2 months prior to VPI repair.

Based on the patient's preoperative nasopharyngoscopy, which noted a central gap on sagittal closure, pharyngeal flap surgery was expected to provide increased tissue in the sagittal place of the nasopharynx and improvement in her VPI. The resulting nasal ports could be closed with considerably less sagittal movement of the lateral pharyngeal wall during swallowing and speech. The patient underwent a superior-based pharyngeal flap in the operating room. General endotracheal (ET) anesthesia was induced per the pediatric anesthesia team. The technique was, briefly, as follows: two vertical incisions were made in the posterior pharyngeal wall from the level of C1 to the larynx. They were connected with a horizontal incision at the larynx and the flap was elevated in the prevertebral plane. The flap was inset into the soft palate and the flap donor site was partially closed with vicryl interrupted sutures and over-sewn uvula. The nasopharyngeal ports were noted to be 4×5 mm bilaterally and patent. The patient was extubated without difficulty but did require positive pressure mask ventilation after ET tube removal to maintain adequate oxygen saturations. The procedure was without intraoperative complication and the patient was transferred to the pediatric ICU (PICU) for recovery with a tongue stay suture in place and on room air.

In the PICU, the patient received IV clindamycin 10 mg/kg q8 h, dexamethasone 1 mg/kg q8 h, and morphine 0.1 mg/kg q2 h as needed. Within hours of arrival to the PICU, she began having desaturations to the high 70 s. She had difficulty tolerating her oral secretions and required frequent suctioning. Chest X-Ray (CXR) demonstrated mild pneumopericardium (Fig. 1). She was placed on a humidified face tent with O_2 4 l/min and a precedex drip was started per the PICU staff. On the morning of post-operative day (POD) #1, the patient was tachypneic to >60 breaths/min. She received respiratory treatments but her saturations and respiratory rate remained labile; her oxygen requirement continued to increase.

Early in the morning of POD#2, the patient became increasingly lethargic with increased work of breathing and desaturations to the 70 s. CXR revealed cervical emphysema, pneumomediastinum, pneumocardium, small bilateral pneumothoraces, and bilateral airspace disease (Fig. 2). Cardiopulmonary resuscitation was started. Her oxygen saturations did not improve despite positive pressure mask ventilation. The decision was made to reintubate the patient as she continued to deteriorate through the resuscitative efforts. Maximal ventilator settings (FiO_2 100%, postitive end-expiratory pressure (PEEP) 8, peak pressure 30 s) yielded oxygen saturations in the 80 s. Flexible bronchoscopy was performed via the endotracheal tube by pediatric otolaryngology; mucus plugging was not identified and removal of ventilator support caused the patient to rapidly desaturate to the 40 s. A dopamine drip was started for hypotension. Pediatric cardiology was consulted and an echocardiogram revealed excellent cardiac function without evidence of tamponade physiology. Pediatric surgery placed a left-sided chest tube to relieve the pneumothorax.

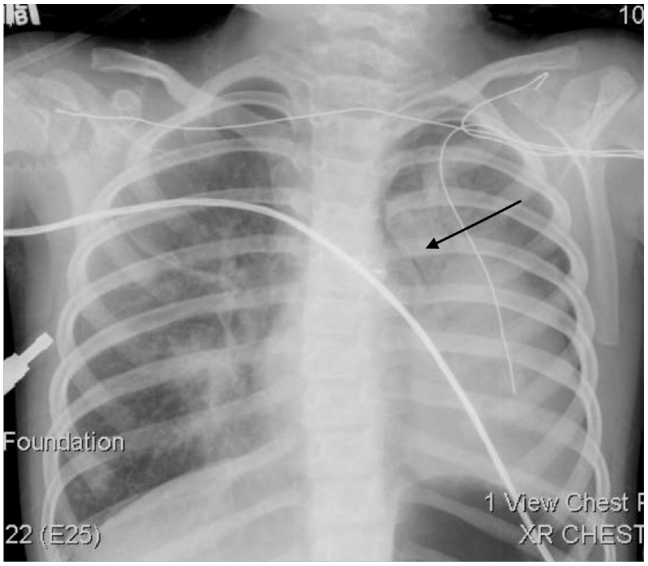

Fig. 1. Plain anterior/posterior (AP) CXR. Arrow indicates mild pneumomediastinum and pneumopericardium. The CXR also demonstrates opacification in the left lower and middle lobes and right lung base.

After these interventions, the patient's oxygen saturations improved to >90% and she tolerated weaning of ventilator settings.

The patient continued to require dopamine infusion on POD#3 for hypotension and bradycardia. She continued to have desaturations despite FiO_2 80% and gentamycin was added to her therapeutic regimen for radiographic evidence of aspiration. An orogastric tube was placed easily by pediatric otolaryngology and enteral feeds were initiated. The patient required placement of a second chest tube on POD#4 for new development of right-sided pneumothorax associated with worsening cervical subcutaneous emphysema. Her respiratory status subsequently improved and

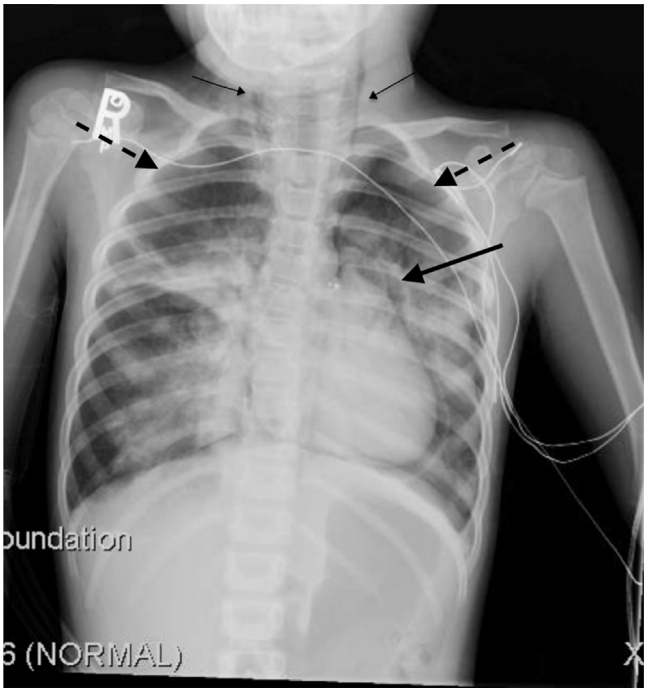

Fig. 2. Plain AP CXR Thick arrow indicates pneumomediastinum and pneumopericardium. Dashed arrows indicate small bilateral pneumothoraces. Fine arrows indicate subcutaneous emphysema in the supraclavicular soft tissues of the neck bilaterally.

she was weaned from pressors. Curiously, her pneumomediastinum and subcutaneous emphysema improved with chest tube placement. Her ventilator settings, particularly her inspiratory time and PEEP, were decreased to avoid further barotrauma. She remained intubated until POD#8 while she was treated for aspiration pneumonia and pseudomonas urinary tract infection. She was able to tolerate oral feedings POD#9 and her chest tubes were removed POD#10. Supplemental oxygen was discontinued on POD#11. She was transferred out of the PICU on POD#12 and discharged to home in good condition on POD#13.

3. Discussion

Here we report an unexpected, severe complication that occurred shortly after superior-based pharyngeal flap surgery for VPI; the etiology of which has not been previously reported in the literature. During the patient's treatment course, multiple interdisciplinary team meetings took place, including pediatric faculty in otolaryngology, intensive care, anesthesia, cardiology, and infectious disease. The suspected etiology of the patient's respiratory failure was air tracking though the visceral or prevertebral fascia into the common pleural spaces of the mediastinum and thorax during the positive pressure mask ventilation after the patient was extubated in the operating room. When she began to decompensate on POD#2, she was again masked with positive pressure prior to intubation. She continued to deteriorate through the resuscitative attempts. Per routine, the flap donor site in the posterior pharyngeal wall was only partially closed with interrupted sutures and over-sewn uvula. A portion of the donor site was left to heal by secondary intention, leaving a port of entry for air to tract through the deep neck fascial planes to the mediastinum. A cause thought to be less likely by our team was mucus-plugging in the tracheobronchial tree leading to auto-PEEP and subsequent air leak into the pleural space. No evidence of mucus plug or tracheobronchial injury was ever observed on flexible bronchoscopy and there was no difficulty with ventilation intraoperatively. Additionally, the initial manifestation of pneumomediastinum/pneumopericardium, as well as bilateral pneumothoraces, makes this etiology less likely. The common pleural cavity in the chest facilitated the spread of air from the deep neck to the mediastinum, pericardium, and pleural spaces of the lungs in our patient.

Pneumomediastinum is thought to be caused by one of four mechanisms: (1) direct air leak from the trachea, bronchus, or esophagus; (2) perforation of a hollow abdominal viscus; (3) air tracking via the fascial planes of the neck; (4) tears in the pulmonary parenchyma with perivascular air tracking known as the Macklin effect [6]. There have been rare cases of traumatic or spontaneous pneumomediastinum reported in the pediatric population [7,8]. There is a bimodal incidence with peaks at less than age 4 years and over age 15 years. The etiology in the younger population is thought to be air leak due to elevated intrathoracic pressure during forceful Valsalva maneuver (coughing, emesis, straining) [7]. Spontaneous pneumomediastinum in children is typically managed conservatively [7,8]. Our patient, with her associated pneumopericardium, bilateral severe pneumothoraces requiring thoracotomy tubes, and cervical subcutaneous emphysema makes this case particularly unusual.

Our patient underwent our standard, detailed preoperative evaluation. Her genetic work up did not reveal a known syndromic association, but our patient did have a history of cardiac defects including VSD and PDA. Her PDA was coiled in a cardiac cath lab and her VSD closed spontaneously, so she had not had any cardiothoracic procedures that may have contributed to her development of this complication. When planning future pharyngeal flaps, pre-operative discussions with pediatric anesthesia will include strict avoidance of high-pressure bag-mask ventilation post-operatively to prevent cervical air tracking.

4. Conclusion

We present the first case of respiratory failure secondary to pneumomediastinum, pneumopericardium, bilateral pneumothoraces, and subcutaneous emphysema following superiorly-based pharyngeal flap for velopharyngeal insufficiency. This is an important consideration that should guide discussions with anesthesia to avoid bag-mask ventilation in the presence of an open surgical wound in the prevertebral fascia. Management is aggressive cardiopulmonary support.

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Conflict of interest statement

None.

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