Delayed diagnosis of bilateral pneumothoraces after a difficult tracheostomy in a 6 week-old with CHARGE syndrome

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ABSTRACT BODY: Introduction: Intraoperative complications of tracheostomy including hemorrhage, perforations of the walls of the trachea and esophagus, tracheocutaneous fistulas, pneumothorax and pneumomediastinum, recurrent laryngeal nerve injury, intraoperative fire are well described in the literature. Bilateral pneumothoraces is an extremely rare complication of tracheostomy. We present the case of a patient with Charge syndrome who required a tracheostomy, complicated by bilateral pneumothoraces.

Case Report: A full-term 6 week-old 2.9 kg girl with CHARGE syndrome (with bilateral choanal atresia) was scheduled for tracheostomy. The baby was initially intubated at birth for respiratory distress and subsequently remained intubated for respiratory failure. In addition to bilateral choanal atresia and coloboma of the eye she was noted to have the dysmorphic features associated with CHARGE syndrome (hypertelorism, low set ears, typically asymmetric square face, malar flattening, and micrognathia), She had previously undergone a PDA ligation.

In the operating room, the patient was positioned on a shoulder roll with the neck fully extended to improve operative exposure. Anesthesia was maintained with sevoflurane 2.5% in oxygen and fentanyl 2 mcg/kg. Spontaneous ventilation was maintained. The surgeon made with a 1-cm vertical incision in the skin overlying the second and the third tracheal rings, followed by a tracheal incision. After the endotracheal tube had been withdrawn 1 cm by the anesthesia team, the surgeon inserted a 3.5 tracheostomy tube into the trachea. The anesthesia machine breathing circuit was connected to the tracheostomy tube but no end-tidal CO$_2$ was recorded after 3 gentle squeezes of the ventilation bag. A change in the ‘feel’ with manual ventilation was also noted by the anesthesiologist and the tracheostomy tube was rapidly removed. Adequate ventilation and normal respiratory pressures were re-established with advancement of the endotracheal tube past the tracheal incision. A second attempt at placing the tracheostomy tube through the same tracheal incision produced a similar result. Suspicion arose that the tip of the tracheostomy tube was abutting the posterior wall of the trachea and impeding passage of air. The shoulder roll was therefore removed and the neck was slightly flexed. The tracheostomy tube was then re-inserted and a normal end-tidal CO$_2$ waveform was obtained. The endotracheal tube was removed and the patient continued to breathe spontaneously through the tracheostomy tube for the remainder of the case. No obvious changes in the dynamics of the respiration or oxygen saturation were noted and the case ended uneventfully.

The patient was transported to the NICU while receiving manual positive pressure ventilation. Upon placement of the patient on the ventilator in the NICU, high peak airway pressures were noted in conjunction with hypotension and a reduced arterial saturation. A stat chest radiograph was obtained that demonstrated bilateral tension pneumothoraces. Bilateral needle decompression was performed with a rush of air, and O$_2$ saturation normalized. No thoracostomy tube was placed and the patient was closely monitored. Serial chest radiographs demonstrated no recurrence of the pneumothoraces. The patient recovered uneventfully.

Discussion: Tracheostomy in children can be associated with significant morbidity and mortality. Most of the children who require tracheostomy have associated congenital or genetic abnormalities. The incidence of complications associated with tracheostomy has decreased over time due to improved intraoperative and postoperative care, however complications still occur. Pneumothorax as the result of a
number of mechanisms is a known complication of difficult tracheostomies. Pneumothorax may result from direct injury to pleura and children are at particular risk because the pleural domes are higher in the neck. Air dissection through the deep layer of the middle cervical fascia can lead to pneumomediastinum and subsequent pneumothorax if the air ruptures into the mediastinal pleura. It is also possible that perforation outside of the lumen of the trachea can occur leading to a dissecting pneumothorax. Difficult surgical exposure may place children with congenital anomalies of the head and neck at greater risk for pneumothorax via these mechanisms. The precise mechanism of injury in this patient is unclear but the diagnosis was delayed by persistence of spontaneous ventilation.

**Conclusion:** In summary, a case of bilateral pneumothoraces as a complication of a tracheostomy is presented. The clinical suspicion for complications should be high when surgical placement is difficult, especially in children with associated congenital anomalies. In this patient maintenance of spontaneous ventilation delayed the diagnosis of pneumothorax. Initiation of positive pressure ventilation after placement of the tracheostomy would have presumably lead to development of the pneumothoraces in the operating room thereby avoiding a crisis situation upon arrival in the NICU. Placement of the thoracostomy tubes is not always necessary for treatment of pneumothoraces in neonates and needle decompression may be sufficient.