

Navigating Complex Airway Management in Status Asthmaticus with Klippel-Feil Syndrome

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Clinical Presentation

A 17-year-old Male with a history of asthma, scoliosis, and Klippel-Feil Syndrome presented to the ED for a one-day history of chest tightness and shortness of breath. The patient’s home asthma control regimen includes Symbicort, Montelukast, and albuterol. Additional history was unable to be identified at this time. Initial ED management included ipratropium-albuterol nebulizers, corticosteroids, and magnesium sulfate, without resolution of symptoms. This prompted admission to PICU for status asthmaticus. Five days later, the patient complained of chest tightness and dyspnea again and was treated with oxygen, nebulized albuterol, systemic glucocorticoids, magnesium sulfate drip, terbutaline, and IV fluids. Rapid Response Team (RRT) was called after one hour secondary to respiratory distress and hypercapnic narcosis (Pre-intubation ABG revealed pH 7.07, pCO2 111.6, pO2 242. HCO3 32.6, SpO2 99%). Due to the patient’s history, he was taken to the OR for an anticipated difficult intubation. Physical examination revealed marked respiratory distress with 70% SpO2 on room air. He was lethargic, utilizing accessory muscles of respiration, and had a poor response to pain stimuli. Diffuse bilateral wheezing and tachycardia were noted. Musculoskeletal examination revealed decreased neck mobility, thoracic scoliosis and kyphosis, and rotational deviation of the thoracic spine. Due to the patient’s abnormal oral and cervical anatomy and limited cervical range of motion, Mallampati score and thyromental distance were unable to be assessed. Otherwise, examination findings were within normal limits.

Anesthesia Course

Endotracheal intubation was indicated for airway protection. Neutral supine cervical spine position was maintained due to limited range of motion. An oropharyngeal airway was placed, and the patient was preoxygenated. Three attempts at intubation were performed by different physicians. The first and second attempts using McGrath and fiberoptic laryngoscopy, respectively, with midline approaches were unsuccessful. The third attempt using GlideScope laryngoscopy was successful to insert a 6.5 mm ETT with visualization of the epiglottis positioned left of midline. Bilateral breath sounds and proper EtCO2 return on ventilator were confirmed. Throughout the procedure, the patient received midazolam, ketamine, succinylcholine, rocuronium, propofol, dexmedetomidine, epinephrine, albuterol, glycopyrrolate, and Lactated Ringers solution. Chest XR confirmed ETT placement 2 cm above the carina. The ventilator was set to a tidal volume (TV) of 400 mL, respiratory rate (RR) of 22/min, fraction of inspired oxygen (FIO2) 50%, and peak end-expiratory pressure (PEEP) of 5 cm H2O. Post-intubation arterial blood gas one hour later revealed pH 7.10, pCO2 91.3, pO2 153, HCO3 28.6, SpO2 98%, Lactate 1.27.

Once stabilized, retrospective record review revealed a history of Tetralogy of Fallot repair with residual pulmonary insufficiency, occiput-C5 cervical fusion, and bilateral orchiopexy. Prior records for status asthmaticus from an outside facility revealed a history of difficult intubations requiring multiple attempts. The first attempt was unsuccessful with asleep fiberoptic intubation. The second attempt utilizing McGrath laryngoscopy was unsuccessful but the third attempt utilizing C-MAC video laryngoscopy at a leftward angle was successful.

The following day, an air leak in the ETT cuff which was confirmed with video laryngoscopy. A fiberoptic bronchoscope was inserted through the ETT with resistance and was unable to advance. Video laryngoscopy revealed the ETT was dislodged. A new 6.0 ETT was inserted at an angle left of midline successfully in one attempt.

Klippel-Feil Syndrome

Klippel-Feil Syndrome (KFS) is a rare bone disorder linked to GDF6, GDF3, or MEOX1 gene mutations, characterized by abnormal fusion of 2+ vertebrae in the cervical spine. Patients classically present with a triad of a short neck, appearance of a low posterior hairline, and decreased cervical spine mobility. KFS is commonly associated with fusions in the thoracic spine causing scoliosis and/or kyphosis, restricting lung mechanics and predisposing patients to spinal stenosis and neurological deficits. Associated cardiac, renal, and GI deformities can be evaluated with ultrasonography., while X-ray and CT are used to assess the extent of spinal fusion or presence of scoliosis. MRI is utilized for patients experiencing neurological deficits. Management can include nonoperative approaches such as physical therapy, or surgery for patients with more spinal instability.

There are multiple considerations related to anesthesia with KFS patients. The short thyromental distance and decreased cervical spine mobility make positioning and airway access considerably more difficult. Restrictive elements from possible scoliosis, kyphosis, and additional rotational components as seen in this patient complicates optimal lung ventilation. Anatomical anomalies such as airway deviation from midline can complicate intubation technique especially under emergent conditions.

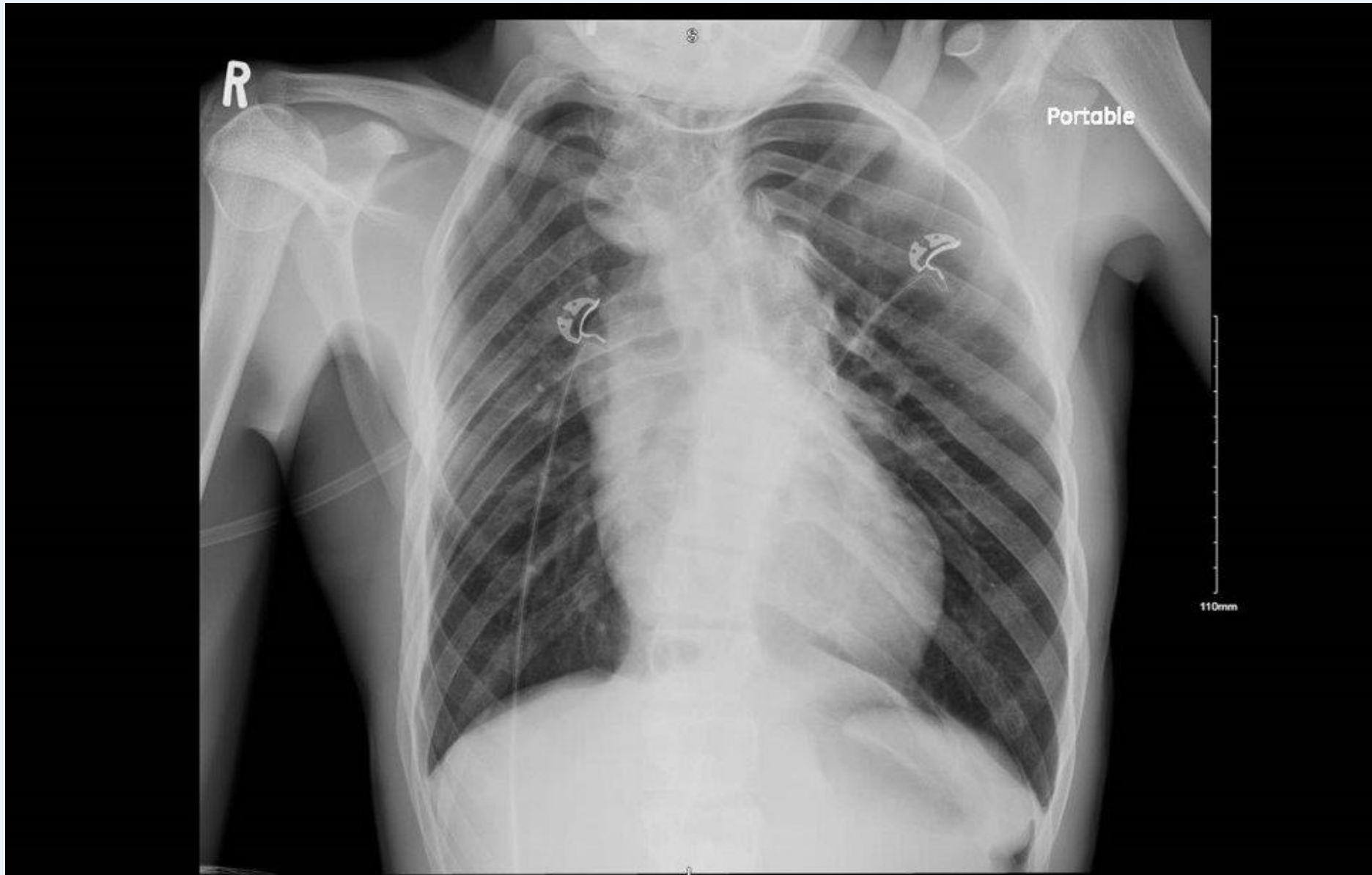


Figure 1: Pre-intubation AP Chest XR

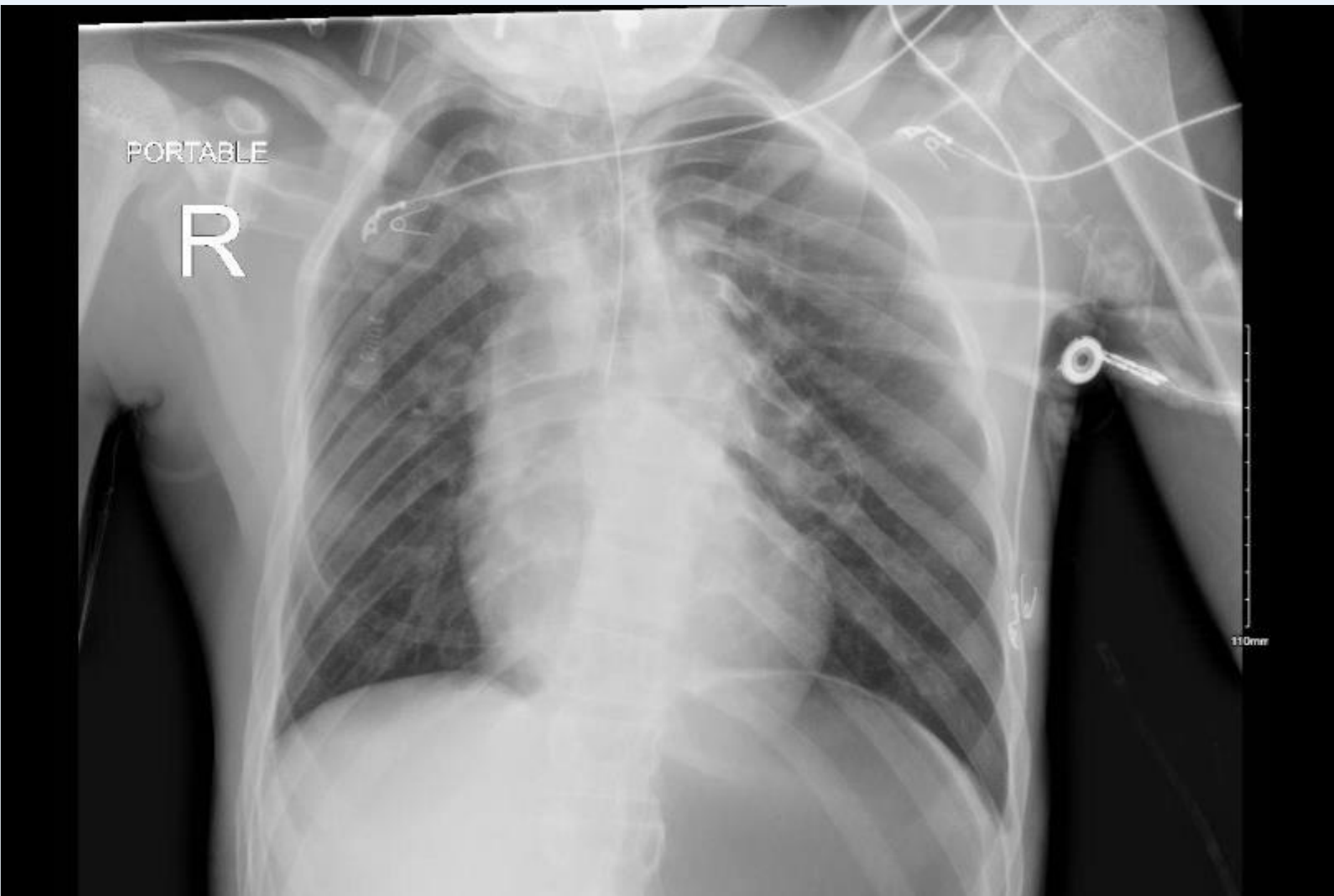


Figure 2: Post-intubation AP Chest XR

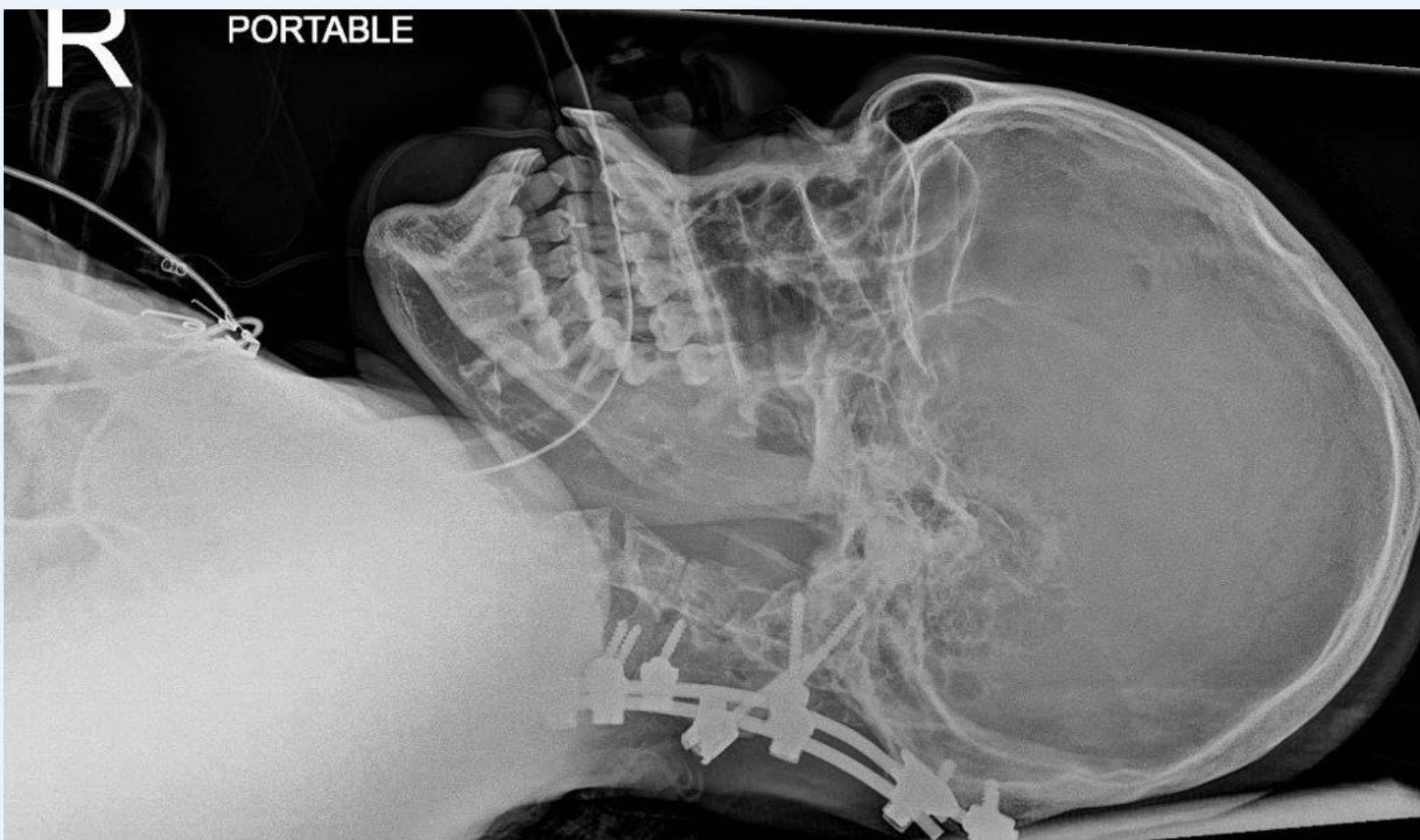


Figure 3: Post-intubation lateral head and neck XR

Status Asthmaticus

Status asthmaticus, a life-threatening medical emergency, is a severe form of asthma exacerbation that is unresponsive to standard treatment protocol. Physiologically, an acute exacerbation occurs when there is severe bronchoconstriction, airway inflammation and hyperreactivity, immune cell infiltration and excess mucus production. While asthma is typically characterized by dyspnea and wheezing, status asthmaticus is characterized by hypoxemia, hypercapnia, and respiratory failure. Both conditions exhibit obstructive lung mechanics. Classically tachycardia, tachypnea, dyspnea, accessory muscle use, and altered mental status are seen on physical examination which should also include vital sign and mental status monitoring, measurement of Peak Expiratory Flow Rate (PEFR) and Forced Expiratory Volume in 1 second (FEV1), measurement of arterial blood gas (ABG), complete blood count (CBC), and basic metabolic panel (BMP).

Status asthmaticus management includes supplemental O2 with target SpO2>92%, IV placement, and initial pharmacologic treatment with continuous nebulized B2 agonists (Ex. Albuterol) for up to 15 mg/hr, and IV corticosteroids (Ex. Methylprednisolone) 2 mg/kg up to 125 mg, then at 0.5-1 mg/kg every 6 hrs. If unsuccessful, add IV magnesium sulfate, epinephrine, and terbutaline while continuously reassessing the need for endotracheal intubation. Consider noninvasive ventilation and administration of ketamine or nitric oxide as continued measures. Further management would include rapid sequence intubation using ketamine and succinylcholine for induction. Propofol may also be utilized as an anesthetic agent given its minor bronchodilatory effects, however in pediatric populations ketamine is preferred. Additional considerations include methylxanthines, IV fluids, antibiotics, mucolytics, and bronchoscopy. Indications for intubation include obtunded mental status, evidence of respiratory fatigue, persistent hypoxia despite multimodal treatment, increasing accessory muscle use or speech changes, decreasing respiratory rate and depth.

Discussion

Patients presenting in status asthmaticus complicate ventilation management with an underlying obstructive element to their lung mechanics which, paired with any alteration in mental status, necessitates the use of rapid sequence intubation with special consideration for choice of induction agents. The history of Klippel-Feil Syndrome complicated airway and ventilation management in multiple ways. The degree of scoliosis, kyphosis, and rotational deviation of the spine leaves his airway positioned left of midline while the patient is in neutral supine positioning, causing difficulty for the physician attempting intubation with a traditional midline approach. Additional anatomical difficulties included the considerably short thyromental distance and limited cervical range of motion, effectively eliminating the possibility of extending the head at the neck for optimal positioning. Thus, lung mechanics are subject to a restrictive element as well. Lastly, the importance of a thorough pre-operative H&P with any possible collateral information is highlighted in this case. Though difficult to obtain under emergent conditions, knowledge of prior history, such as the successful leftward-angled approach, can influence approach to intubation. Awareness of Klippel-Feil Syndrome and its potential complications in airway and anesthetic management, can help improve quality of care and patient outcomes especially under emergent settings.

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