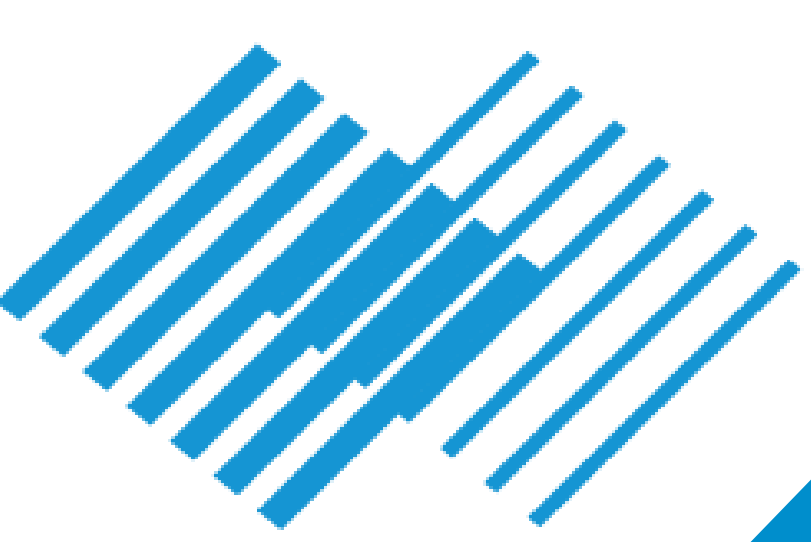




Anesthetic Management of an Infant with Trisomy 14 Undergoing Tracheostomy Placement

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BACKGROUND

- Trisomy 14 is a rare congenital disorder with only a few hundred reported cases around the world
- It is associated with developmental and craniofacial abnormalities, including growth and psychomotor restriction, broad nose, micrognathia, cleft palate, highly arched palate, short neck, and congenital heart disease
- Almost all reported cases involve mosaicism, as complete trisomy 14 is highly lethal in utero

PMH

- Our patient was a 5-month old ex-34-weeker weighing 3.7kg who presented for tracheostomy placement
- Shortly after birth, he had a 3-month stay in the NICU
- Past medical history was significant for mosaic trisomy 14, laryngomalacia, chronic lung disease with baseline SpO2 87%, hypotonia, PFO and failure to thrive requiring a G-tube

HPI

- He was admitted to the PICU for acute respiratory failure due to viral bronchiolitis
- Required nightly CPAP and daytime nasal cannula, supplemented with nebulizers
- Given his dysmorphic facial features, ENT was consulted and performed direct laryngoscopy and rigid bronchoscopy with an anterior commissure laryngoscope
- Findings were notable for micrognathia, retrognathia, nasal stenosis, laryngomalacia, and omega-shaped epiglottis with a grade 2-3 view of vocal cords
- Consented for tracheostomy with consideration for future supraglottoplasty

INDUCTION

- Pt arrived in OR on room air in no distress with a 24g IV in the L hand
- He was placed on standard ASA monitors, including EKG, BP cuff on the R arm, and pulse oximetry on the L foot, with a backup pulse oximeter on the contralateral foot
- Due to the tenuous airway, the anesthesia and ENT teams jointly agreed on an awake intubation to minimize the risk of respiratory compromise

INTUBATION

- After preoxygenation, the anesthesia team proceeded with the awake intubation with a size 3.5 oral rae tube using a CMAC and a MAC #0 video laryngoscopy blade, revealing a transient view of the arytenoids and a grade I view of the vocal cords
- Upon attempts to pass the tube through the cords, Pt frequently coughed and subsequently desaturated, and he subsequently improved when the tube was withdrawn
- Because he was unable to tolerate awake intubation, the decision was made to fully induce the patient for optimal conditions for intubation using 5mg propofol and 3mg rocuronium
- Patient was intubated successfully by the surgeon via rigid telescope with an anterior commissure laryngoscope
- Placement was confirmed with end-tidal CO2, and the tube was secured at a depth of 11cm.

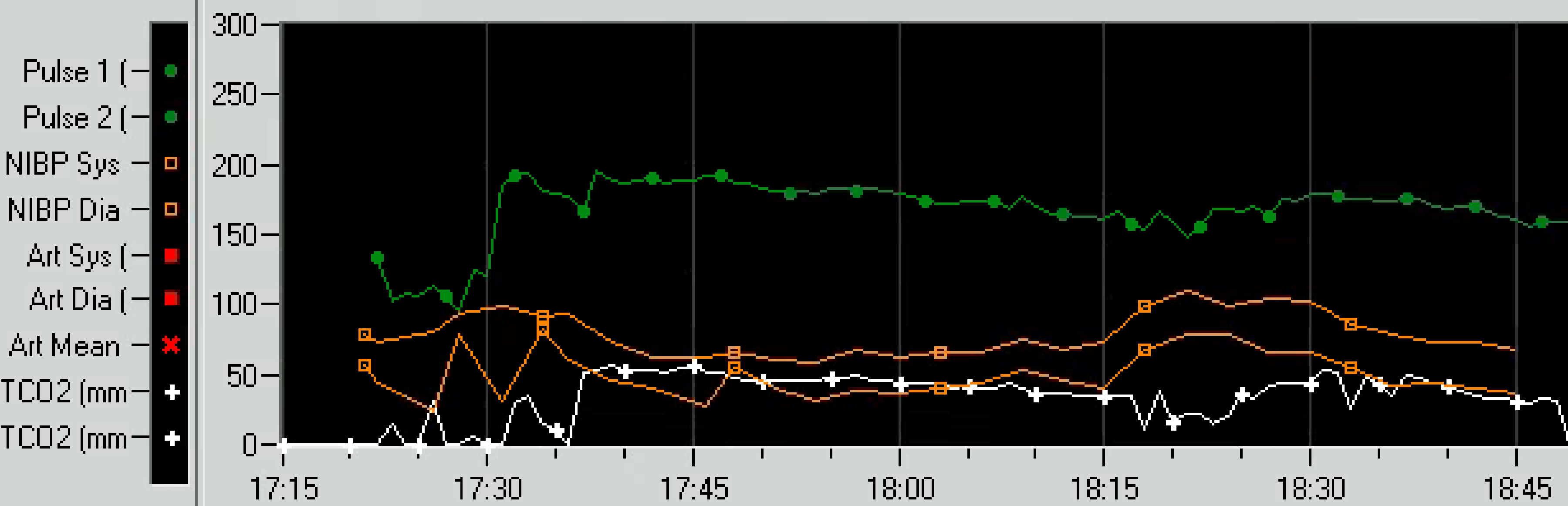


Figure 1: Vital signs throughout procedure

MAINTENANCE

- Surgical anesthesia was maintained with an additional 10mg propofol, 5mcg fentanyl, and 1% sevoflurane
- During placement of the size 3.5 Shiley tracheostomy tube, the anesthesia team withdrew the oral rae tube until slightly above the vocal cords
- After successful placement of the tracheostomy tube, the endotracheal tube was removed, and the patient was successfully ventilated via the newly-placed tracheostomy

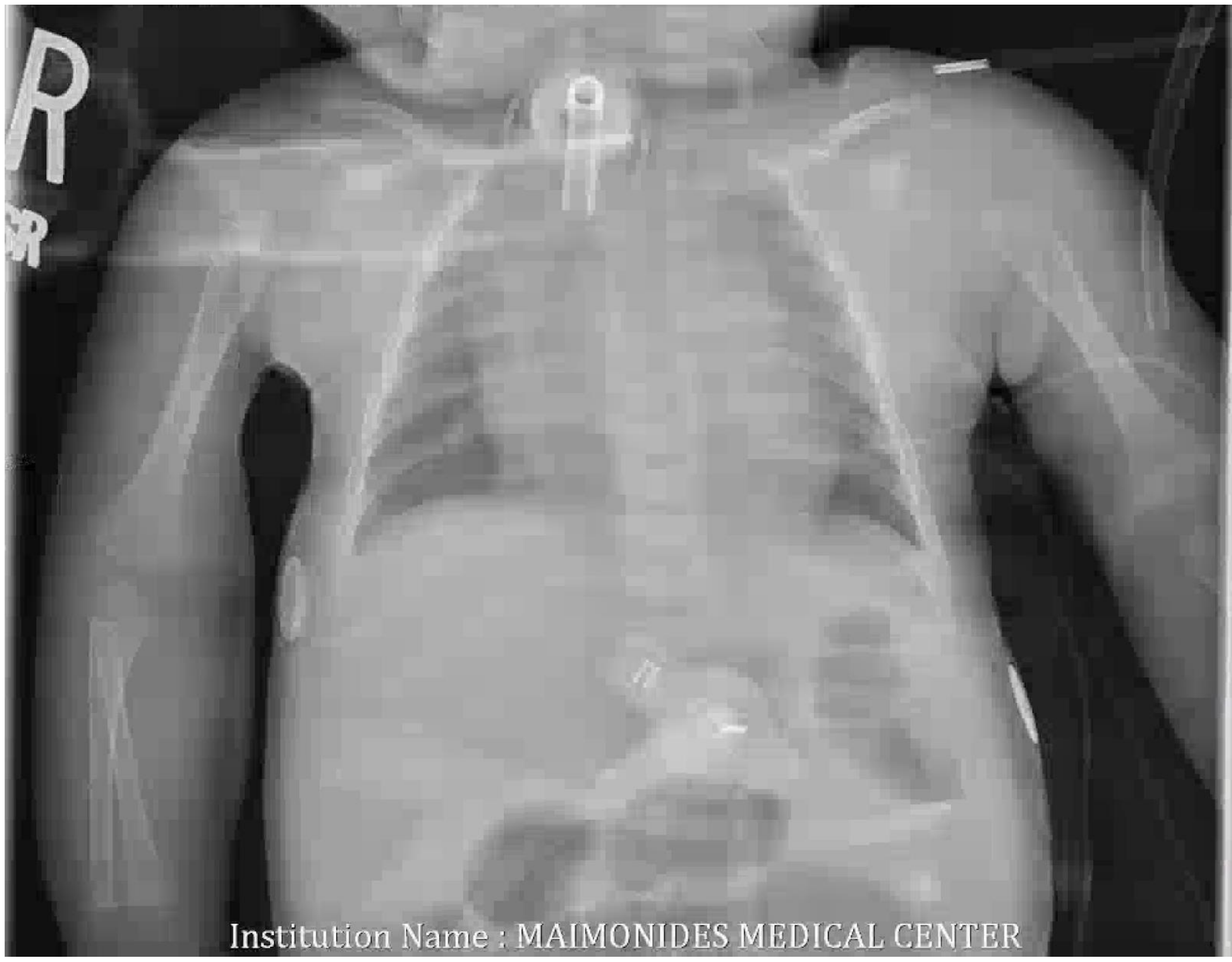


Figure 2: Post-operative chest x-ray, showing new tracheostomy

CONSIDERATIONS

- Due to the high incidence of craniofacial abnormalities in patients with trisomy 14, anesthesia providers must anticipate and manage these difficult airways
- Careful history must also be taken to elucidate any congenital cardiac abnormalities