

Airway Maintenance in a Newborn with Hanhart Syndrome

A.V. Mostovoy and K.D. Gorelick

Children's Hospital No. 1, St. Petersburg, Russia

Described more than half a century ago, Hanhart syndrome is a condition characterized by hypoplasia of the tongue and lips, and other oral-facial anomalies. In most cases, inheritance is autosomal dominant and all reported cases are born to consanguinity related parents. To our knowledge, full fusion of upper and lower jaws has not been reported as part of the syndrome. We report such a case.

A male full term appropriate for gestational age infant was born via c-section with Apgar score 6/6. Mother is 25-year-old Gravida IV Para II (one full-term healthy infant, two induced abortions), parents are first cousins. Initial prenatal ultrasound (late prenatal care at 26 weeks) did not reveal any abnormalities except for polyhydramnios.

Since birth - serious condition secondary to multiple congenital anomalies, including complete mandibular-maxillary fusion (no oral opening was seen), fusion of the lips, significant mandibular hypoplasia. Congenital anomalies also included short neck, small eye openings, camptodactyilia, hypogonadism with bilateral cryptorchidism, wide spaced nipples. The child was intubated intranasally with a size 2 tube to the oropharynx in the neonatal intensive care unit, and assisted ventilation was initiated to alleviate respiratory distress.

Positive findings on CT revealed gross defects of the facial bones combined with a hypoplastic lower jaw and a possibly complete fusion of the processus alveolaris of the lower and upper jaws with the lower alveolar bone located behind the upper one; unidentifiable temporomandibular joints, normal tongue lodged in the hypoplastic oral cavity; normal choanae in slit-like naso-pharynx; narrowed proximal trachea below the C4 box, irregularly shaped axial cut and dysplastic bones at the base of the skull.

Difficult intubations are rarely encountered in neonatal practice. This tracheal intubation required a very small bronchoscope (2 mm in diameter), which we did not have at our institution. At the same time intubation with a standard bronchoscope was technically impossible. While attempting a tracheostomy to open the child's airways for mechanical ventilation the tracheostomy tube entered the mediastinum because of the narrowness of the patient's trachea. The subsequent acute pneumo-mediastinum led to the patient's death.

In this case, the attempt at a tracheostomy was born of desperation.



Dr. Mostovoy