International Journal of Pediatric Otorhinolaryngology 78 (2014) 1799–1803

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Case Report

Resolution of airflow obstruction on polysomnography after laryngotracheal reconstruction with anterior tracheal wall suspension in a patient with DiGeorge Syndrome

Cindy Jon [a,b,*], Sarah E. Mitchell [b,1], Ricardo A. Mosquera [a,2], James M. Stark [a,3], Sancak Yuksel [c,4]

[a] Department of Pediatrics, University of Texas Health Science Center, Houston, TX, USA
[b] Memorial Hermann Memorial City Hospital, Pediatric Sleep Center, Houston, TX, USA
[c] Department of Otorhinolaryngology—Head & Neck Surgery, University of Texas Health Science Center, Houston, TX, USA

ARTICLE INFO

Article history:
Received 6 May 2014
Received in revised form 22 July 2014
Accepted 24 July 2014
Available online 2 August 2014

Keywords:
Obstructive sleep apnea
DiGeorge Syndrome
Sleep disordered breathing
Anterior cricoid suspension
Anterior tracheal wall suspension

ABSTRACT

DiGeorge Syndrome (DGS) may be associated with airway abnormalities including laryngomalacia and suprastomal collapse of the trachea (SCT), which may lead to sleep disordered breathing (SDB). We present a 4-year-old boy with DGS, SCT, and SDB by polysomnography (PSG) while the tracheostomy tube was capped. The patient underwent anterior tracheal wall suspension (ATWS) with concurrent tracheostomy decannulation. Following the repair, the patient experienced improved airway patency visually and by PSG with resolution of obstructive sleep apnea and hypoventilation. ATWS is an effective method to repair SCT in selected patients and may lead to early decannulation and improvement of SDB.

1. Introduction

DiGeorge Syndrome (DGS, Chromosome 22q11.2 deletion syndrome) is a congenital disorder that affects multiple organ systems with variability in presentation and severity. Characteristic features of DGS include hypoplasia or aplasia of the thymus and/or the parathyroid glands, congenital heart defects, facial dysmorphism, structural airway anomalies, ear abnormalities, gastro-esophageal reflux (GER), learning disabilities, and/or immunodeficiency [1]. Commonly reported airway abnormalities in DGS include cleft lip and palate, tracheoesophageal fistula, esophageal atresia, choanal atresia, reduction in the number of tracheal rings, shortened trachea, tracheal compression, small thyroid cartilages, laryngomalacia, tracheomalacia, bronchomalacia, and tracheal bronchus [2]. DGS is linked to defective structures that arise from the third and fourth embryonic pharyngeal arches due to defects in chromosome 22. DGS has been associated with the micro deletion of band 11 on chromosome 22 and as such is also referred to as 22q11 deletion syndrome [3].

Sleep-disordered breathing (SDB) is characterized by disrupted airflow during sleep that may lead to sleep fragmentation and result in cardiovascular, metabolic, neurocognitive, and behavioral impairments. SDB includes obstructive sleep apnea (OSA), central sleep apnea (CSA), hypoventilation, hypoxemia, and primary snoring [4]. SDB results in either partial or complete absence of airflow that disrupts sleep. Diagnostic polysomnography (PSG) determines the presence and severity of SDB. In children with typical SDB symptoms, an apnea hypopnea index (AHI) greater than 1/h is diagnostic for OSA, and a central apnea index (CAI) greater than 3/h is indicative of CSA [5,6]. Hypoventilation is diagnosed if end tidal carbon dioxide ($EtCO_2$) is greater than 50 torr for more than 25% of the total sleep time (TST) [7].

* Corresponding author at: Department of Pediatrics, Pediatric Pulmonology University of Texas Health Sciences Center at Houston 6431 Fannin, MSB 3.228, Houston, TX 77030, USA. Tel.: +1 713 500 5650; fax: +1 713 500 0588.
E-mail addresses: cindy.jon@uth.tmc.edu (C. Jon), sarah.mitchell@memorialhermann.org (S.E. Mitchell), ricardo.a.mosquera@uth.tmc.edu (R.A. Mosquera), james.m.stark@uth.tmc.edu (J.M. Stark), sancak.yuksel@uth.tmc.edu (S. Yuksel).
[1] Tel.: +1 713 242 4447.
[2] Tel.: +1 713 500 5650.
[3] Tel.: +1 713 500 5650.
[4] Tel.: +1 713 500 6000.

http://dx.doi.org/10.1016/j.ijporl.2014.07.035
0165-5876/

The incidence of OSA and hypoventilation at baseline and after surgical airway reconstruction in DGS patients has not been widely reported. We present a child with DGS and suprastomal collapse of the trachea (SCT). Baseline PSG showed significant OSA, sleep hypoventilation, and mild CSA with tracheostomy tube capped. The patient underwent anterior tracheal wall suspension (ATWS) with concurrent tracheostomy decannulation which resulted in resolution of OSA and sleep hypoventilation on the post-operative PSGs.

2. Case report

We present a 4 year-old boy with DGS who was born at 24 weeks gestation with a complicated neonatal intensive care unit (NICU) course. He required a tracheostomy tube placement and prolonged mechanical ventilation for severe laryngomalacia, failure to extubate, and bronchopulmonary dysplasia (BPD). He had patent ductus arteriosus (PDA) ligation at 7 weeks of age. Head ultrasound showed grade II intraventricular hemorrhage (IVH). He had a gastric tube placement and laparoscopic fundoplication for feeding intolerance and GER, respectively.

At 11 months of age, he had mircrodebridement of suprastomal granuloma. By 15 months old, direct laryngoscopy and bronchoscopy showed granulation tissue around the stoma which was revised, moderate to severe suprastomal collapse, minimal granuloma, inflamed tracheobronchial tree and mild tracheobronchomalacia. By 23 months of age, he was successfully weaned off of mechanical ventilator support. At 25 months old, airway endoscopy showed 50% hypertrophic adenoid tissue and 4+ hypertrophic tonsils; moderate-to-severe suprastomal collapse (Fig. 1A and B); no subglottic stenosis or another tracheobronchial lesion; hypertrophic peristomal granulation tissue; and possible hypomobility of the left vocal cord. He underwent an adenotonsillectomy (AT) and tracheostoma revision with flap advancement. Tracheostomy tube capping trials were initiated.

Due to stridor on exam at 28 months of age, the patient was referred to the Memorial Hermann Memorial City Pediatric Sleep Center for a diagnostic PSG with the tracheostomy tube capped prior to consideration for decannulation. PSG with tracheostomy tube capped revealed significant OSA with an overall AHI of 7.1/h, obstructive AHI of 5/h, CAI of 2.1/h, and elevated end tidal CO_2 diagnostic for sleep hypoventilation (Table 1).

At 3 years of age, the patient underwent laryngotracheal reconstruction with ATWS and tracheostomy decannulation (Fig. 2A and B). ATWS is a modification of the anterior cricoid suspension (ACS) procedure first described by Azizkhan et al., in a one stage approach [8]. After direct laryngoscopy and bronchoscopy, horizontal incisions were made in the peristomal area extending 1 cm–1.5 cm laterally in either direction. The incisions were made through the dermis. Bovie cautery was then used to raise sub-platysmal flaps superiorly to the level of the thyroid cartilage and inferiorly down to the sternum. Subsequently, the strap muscles were identified and systematically divided and retracted laterally. This area was scarred from prior tracheotomy. Care was taken to stay in the midline and the surgeon dissected down directly onto the trachea. After the trachea was encountered, the assistant surgeon went to the head of the bed and performed an endoscopy. Under direct visualization (Fig. 3A–C) to directly mark the extent of the collapse, 27-gauge needles were placed through the trachea and then visualized in the endotracheal lumen. The external tracheal areas that marked the extent of the internal flaps were noted and marked, and the anterior suspension of the trachea was performed. This was accomplished by placing 3/0 PDS™ sutures to the right and left of midline vertically at the area of the suprastomal collapse. Because of the cartilage buckling, a vertical incision was made in the midline anterior trachea to correct the defect. This was our modification of the original Azizkhan procedure. It was noted that traction with the previously placed sutures did help reduce the collapse, but not totally. Additional horizontal incisions were placed at the superior and inferior aspects of the incision in the trachea. By systematically visualizing the tracheal lumen directly with bronchoscopy while addressing the tracheal releasing incisions and retraction sutures, the proper configuration for opening of the anterior segment of the trachea was found. After optimization, the sutures on both sides of the tracheal wall were anchored to the strap muscles close to their attachments on the sternum. Proper placement was confirmed by doing a bronchoscopy and observing that the lumen was significantly expanded inside the trachea with the diameter approaching that of a normal airway. Several Vicryl™ stitches were also placed to help reduce some of the peristomal soft tissue that was collapsing through the tracheotomy, and two other PDS sutures were placed from the suprasternal area to the anterior tracheal soft tissues to help secure the plication. Under direct laryngoscopy, the anesthesia endotracheal tube was removed from

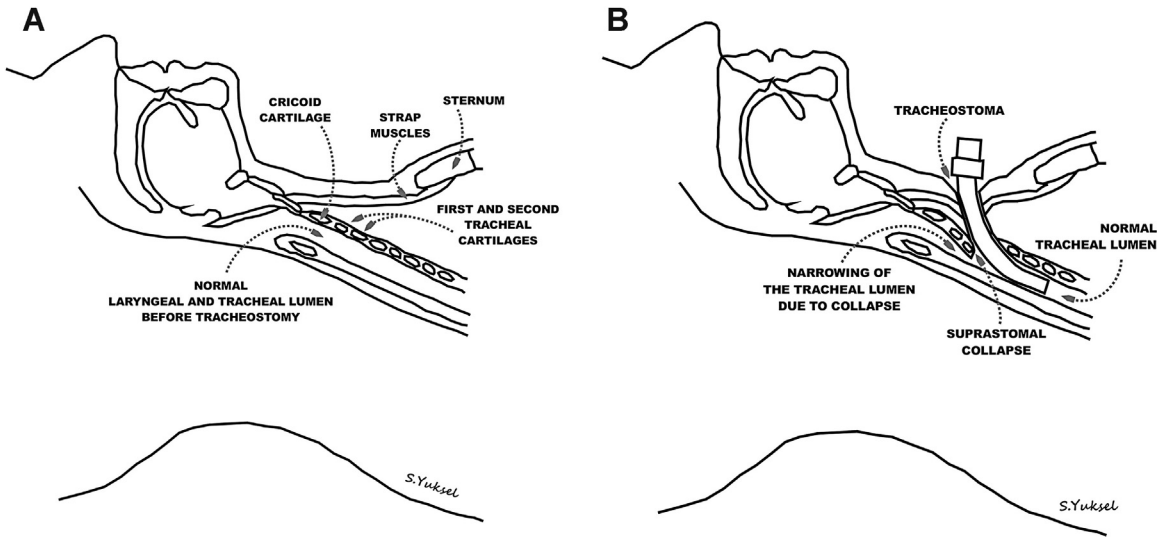

Fig. 1. (A) Normal laryngeal and tracheal anatomy. (B) Suprastomal collapse with tracheostomy tube.