

Congenital High Airway Obstruction Syndrome and Airway Reconstruction

An Evolving Paradigm

Christopher J. Hartnick, MD; Michael Rutter, MD; Florian Lang, MD; J. Paul Willging, MD; Robin T. Cotton, MD

Objectives: To refine the classic definition of, and provide a working definition for, congenital high airway obstruction syndrome (CHAOS) and to discuss the various aspects of long-term airway reconstruction, including the range of laryngeal anomalies and the various techniques for reconstruction.

Design: Retrospective chart review.

Patients: Four children (age range, 2-8 years) with CHAOS who presented to a single tertiary care children's hospital for pediatric airway reconstruction between 1995 and 2000.

Conclusions: To date, CHAOS remains poorly described in the otolaryngologic literature. We propose

the following working definition for pediatric cases of CHAOS: any neonate who needs a surgical airway within 1 hour of birth owing to high upper airway (ie, glottic, subglottic, or upper tracheal) obstruction and who cannot be tracheally intubated other than through a persistent tracheoesophageal fistula. Therefore, CHAOS has 3 possible presentations: (1) complete laryngeal atresia without an esophageal fistula, (2) complete laryngeal atresia with a tracheoesophageal fistula, and (3) near-complete high upper airway obstruction. Management of the airway, particularly in regard to long-term reconstruction, in children with CHAOS is complex and challenging.

Arch Otolaryngol Head Neck Surg. 2002;128:567-570

From the Department of Otolaryngology, Harvard Medical School and Massachusetts Eye and Ear Infirmary, Boston (Dr Hartnick); the Department of Pediatric Otolaryngology, Children's Hospital Medical Center, Cincinnati, Ohio (Drs Rutter, Willging, and Cotton); and the Otorhinolaryngology Service, Centre Hospitalier Universitaire, Lausanne, Switzerland (Dr Lang).

CONGENITAL HIGH airway obstruction syndrome (CHAOS) was defined by Hedrick et al¹ in 1994 as upper airway obstruction that is diagnosed in utero by ultrasound, with concomitant findings of large echogenic lungs, flattened or inverted diaphragms, dilated airways distal to the obstruction, and fetal ascites or hydrops. Of the 4 children with CHAOS who were described by Hedrick and colleagues, none survived past childbirth. Since 1994, there have been several reports of in utero diagnosis of CHAOS, with survival enabled by means of the EXIT (ex utero intrapartum treatment) procedure.² The EXIT procedure was originally designed to treat children with large cervical masses (eg, hemangiomas and teratomas) that were diagnosed in utero and that produced airway obstruction.^{3,4} Of key importance to this type of procedure is the multidisciplinary involvement of the obstetrician, anesthesiologist, otolaryngologist, pediatric surgeon, and neonatologist.

The hallmark findings of CHAOS that were described by Hedrick and coworkers are found consistently only when pre-

natal ultrasonography is performed and, importantly, only when there is complete high upper airway obstruction with no tracheoesophageal connection. The findings relate to the described pathogenesis of the syndrome, whereby the fluid produced by fetal lung tissue has no means of being excreted into the amniotic fluid owing to the upper airway obstruction.¹ The lungs therefore expand and produce the flattening of the diaphragm and the hyperechogenic lung fields that are seen on ultrasound. If the lung fields expand to the point of producing esophageal compression, polyhydramnios may occur as a result of impaired swallowing of amniotic fluid.

The fundamental pathophysiology of CHAOS is altered when there is either an incomplete upper airway obstruction or a tracheoesophageal fistula (TEF), as the fetal lung fluid now has a way to flow from the pulmonary system, and the lung fields therefore do not expand outward. In utero ultrasound may therefore not diagnose the impending danger due to the upper airway obstruction. Survival in such cases therefore depends on the degree of upper airway obstruction, the ability to tra-

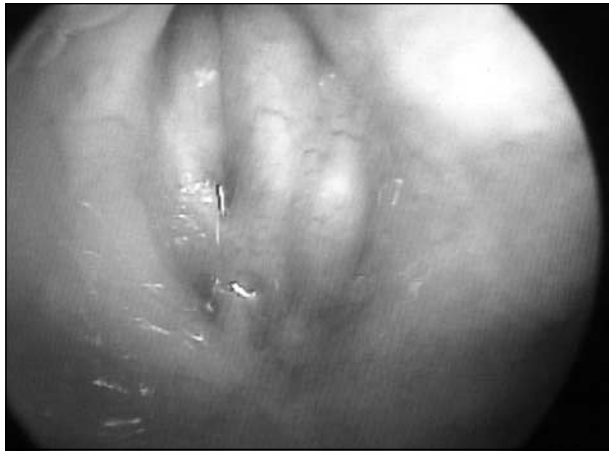


Figure 1. Preoperative endoscopy showing complete laryngeal atresia.



Figure 3. Postoperative view at the time of T tube removal.

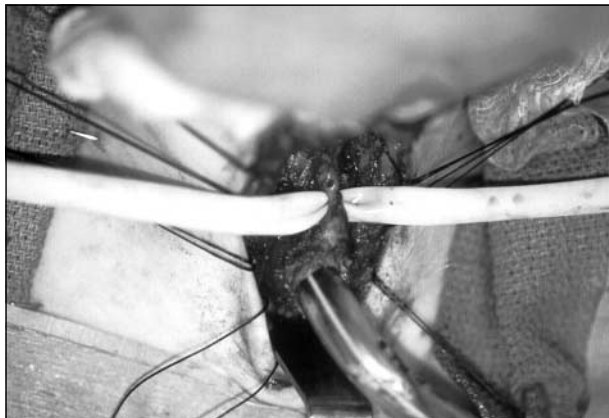


Figure 2. Intraoperative view with a Penrose drain elevating narrow cordlike upper tracheal/subglottic stenosis that extends from the tracheal stoma site to the glottis.

cheally intubate the child through the TEF, or the ability to perform an emergent tracheostomy. Also, regardless of whether the diagnosis was made in utero, management of the survivor's upper airway obstruction is a challenging surgical process. The following 4 cases represent the spectrum of clinical CHAOS and the various modalities used for pediatric airway reconstruction. To our knowledge, this article presents the first description in the otolaryngologic literature of CHAOS and its long-term manifestations.

REPORT OF CASES

CASE 1

A 6-year-old boy was born at an outside institution with complete laryngeal atresia and a TEF. He was intubated emergently through the fistula, and a tracheostomy was subsequently performed. The TEF was then repaired at a later date. Other pertinent findings included an imperforate anus, microgastria, an atrial septal defect, and a ventricular septal defect. At the time of presentation to the Children's Hospital Medical Center, Cincinnati, Ohio, laryngoscopy and bronchoscopy revealed a complete atretic plate extending from the glottis, where

there were no discernible vocal folds to the superior extent of the tracheal stoma site (**Figure 1**). Intraoperative evaluation of the airway revealed a cordlike atretic cricoid cartilage and an upper tracheal segment with a flattened posterior cricoid plate (**Figure 2**). A partial cricotracheal resection with thyrotracheal anastomosis was performed using a T tube for stenting purposes. The T tube has since been removed, and the patient is undergoing plugging and proceeding toward decannulation (**Figure 3**).

CASE 2

A 7-year-old boy was born with stridor and in immediate airway distress requiring emergent tracheotomy. Laryngoscopy and bronchoscopy revealed a tight grade III Myer-Cotton subglottic stenosis with posterior glottic extension. He underwent a tracheostomy immediately after birth. Other physical findings included pectus excavatum. The patient was referred to Children's Hospital Medical Center for evaluation at the age of 3 years, at which point the laryngeal findings were confirmed (**Figure 4**). No vocal folds could be identified. The patient first underwent a laryngofissure with placement of a No. 6 Albuquer above-stoma stent and subsequently underwent a double-stage anterior costal cartilage laryngotracheal reconstruction. His airway currently has a 4.5 endotracheal tube with a 15-cm water leak; he is currently undergoing a plugging trial and moving toward decannulation (**Figure 5**).

CASE 3

A 1-year-old Venezuelan boy underwent a tracheostomy at birth for laryngeal atresia. Other diagnoses included pectus excavatum, renal ptosis, and pulmonary hypoplasia. He was referred to Children's Hospital Medical Center, where laryngoscopy and bronchoscopy revealed a tight grade III subglottic stenosis with congenital lateral and posterior shelves. As the patient had a persistent oxygen requirement, the decision was made to perform airway reconstruction, with the goal of achiev-



Figure 4. Preoperative view showing tight grade III Myer-Cotton subglottic stenosis with narrow posterior subglottic and posterior glottic lumen.

ing a safe airway with delayed decannulation. Therefore, a double-stage posterior costal cartilage laryngo-tracheal reconstruction was performed, with placement of a No. 6 Albuquer above-stoma stent. When the stent was removed, the patient had a widely patent airway. He has returned to Venezuela. The plan is to reevaluate his oxygen requirement in 1 year and to consider decannulation at that point.

CASE 4

A 12-year-old boy underwent an emergent tracheostomy at birth for complete laryngeal atresia. Other findings included bilateral inguinal hernias and prune-belly syndrome. When he was 8 years old, he was seen and evaluated at Children's Hospital Medical Center, where bronchoscopy revealed a complete laryngeal atresia with subglottic and upper tracheal extension (**Figure 6**). His operative course has been complicated: he has undergone a 4-quadrant split laryngotracheal reconstruction, placement of a T tube, and a cricotracheal resection with thyrotracheal anastomosis, and he recently underwent a tracheal homograft placement.

COMMENT

The classic definition of CHAOS as described by Hedrick et al¹ relies on in utero ultrasound for diagnosis and provides a distinct survival advantage to the neonates who are affected because it allows the proper interventional team to be assembled and prepared at the time of delivery. Nevertheless, even given a timely diagnosis and proper management, there have been only a handful of children who have survived. The possibility for survival becomes more dire when ultrasound evaluation is either not performed or nonrevealing. As noted above, the classic findings of CHAOS have been described only in the context of complete upper airway obstruction with no connection to the esophagus. Reports of children born with complete laryngeal atresia with a TEF and without the classic findings of CHAOS have been noted anecdotally by one senior surgeon (R.T.C., oral communication, May 10, 2001). As has

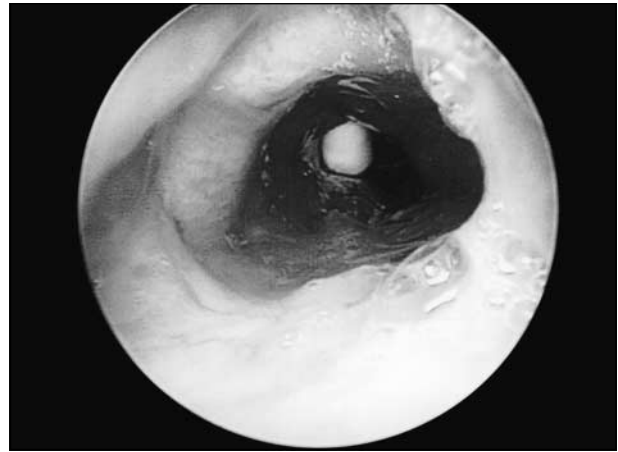


Figure 5. Postoperative view 1 month after T tube removal.



Figure 6. Preoperative endoscopy showing complete laryngeal atresia.

been noted elsewhere, temporary ventilation through this TEF can be lifesaving.⁵ In any case, when there has been no prenatal diagnosis, survival depends upon emergent airway management. The final common pathology is the same: a life-threatening airway obstruction that requires emergent operative intervention. The 4 cases reported herein represent examples of fortunate survivors. The airway management for these children was 2-fold: (1) emergent airway intervention and (2) delayed reconstruction.

As children with complete or near complete laryngeal atresia present with similar symptoms at birth and require a similar treatment course, both immediately and with regard to airway reconstruction, we propose to amend the classic definition of CHAOS to include the various types of high airway obstruction that may be encountered. The amended definition would include any neonate who needs a surgical airway within 1 hour of birth owing to high upper airway (ie, glottic, subglottic, or upper tracheal) obstruction and who cannot be tracheally intubated other than through a persistent TEF. Therefore, CHAOS has 3 possible presentations: (1) complete laryngeal atresia without an esophageal fistula, (2) complete laryngeal atresia with a TEF, and (3) near-complete high upper airway obstruction. One subset of

children who are described by this condition would be identifiable on prenatal ultrasound; the others would not. Children with congenital obstructing masses occluding the airway are excluded from this definition of CHAOS, as they may share a similar common scenario in terms of acute management, but the long-term management of their airway is quite different.

The treatment of children with CHAOS can be divided into immediate and long-term care. As has been noted above, emergent airway management at the time of delivery is enhanced if there has been prenatal diagnosis. A proper team can be assembled, and an EXIT procedure can be planned and carried out. Unfortunately, however, the diagnosis is not always made in utero. Emergent airway management then consists of either tracheoesophageal intubation or emergent tracheostomy. Airway reconstruction is delayed until the child is stabilized and has been allowed to grow.

To our knowledge, there have no reports of subsequent airway reconstructions in children with CHAOS who have survived the perinatal period to date, possibly because there are so few survivors. Pediatric airway reconstruction in this population is challenging. An accurate baseline assessment must be made regarding the child's baseline medical status and comorbid problems, in particular any cardiopulmonary or neurological problems that might preclude decannulation. A careful endoscopy with both flexible and rigid equipment is necessary to assess the mobility of the vocal cords and the degree and length of stenoses. Immobile vocal cords may predispose to an increased risk of postoperative decannulation, and a fiberoptic endoscopic evaluation of swallowing is invaluable in providing information about the child's ability to protect his or her airway or the likelihood of aspiration. Pediatric airway reconstructive surgery should be delayed in the child who is believed to be at risk for aspiration, as opening the glottic and subglottic airway will only increase the likelihood of chronic aspiration and may spur the development of chronic lung disease. Over the past year, 1 child has been evaluated at Children's Hospital Medical Center who was diagnosed with CHAOS in utero, underwent a successful EXIT

procedure, and was under consideration for airway surgery. The patient was believed to be highly likely to have severe aspiration after surgery, and the reconstructive surgery was delayed.

There are a variety of options for surgical reconstruction, including the use of a T tube, cartilage graft reconstruction, cricotracheal resection, and tracheal homografting, and the approach should be individually tailored for each case. The diversity of the possible surgical techniques reflects the diversity that makes up our working definition of CHAOS. Although the final common pathway may produce similar symptoms, the degree of airway obstruction may be near complete or complete, and may involve one discrete level of the airway or may extend from the glottis to the subglottis to the upper trachea. On the other hand, eventual decannulation is a possibility for children with CHAOS, and the families involved should be counseled accordingly.

Accepted for publication August 29, 2001.

Corresponding author and reprints: Christopher J. Hartnick, MD, Department of Otolaryngology, Harvard Medical School, Massachusetts Eye and Ear Infirmary, 243 Charles St, Boston, MA 02114 (e-mail: christopher_hartnick@meci.harvard.edu).

REFERENCES

1. Hedrick MH, Ferro MM, Filly RA, Flake AW, Harrison MR, Adzick NS. Congenital high airway obstruction syndrome (CHAOS): a potential for perinatal intervention. *J Pediatr Surg.* 1994;29:271-274.
2. DeCou JM, Jones DC, Jacobs HD, Touloukian RJ. Successful ex utero intrapartum treatment (EXIT) procedure for congenital high airway obstruction syndrome (CHAOS) owing to laryngeal atresia. *J Pediatr Surg.* 1998;33:1563-1565.
3. Catalano PJ, Urken ML, Alvarez M, et al. New approach to the management of airway obstruction in "high risk" neonates. *Arch Otolaryngol Head Neck Surg.* 1992;118:306-309.
4. Langer JC, Tabb T, Thompson P, Paes BA, Caco CC. Management of prenatally diagnosed tracheal obstruction: access to the airway in utero prior to delivery. *Fetal Diagn Ther.* 1992;7:12-16.
5. Cohen MS, Rothschild MA, Moscosco J, Shlasko E. Perinatal management of unanticipated congenital laryngeal atresia. *Arch Otolaryngol Head Neck Surg.* 1998; 12:1368-1371.