Laryngomalacia: patient outcomes following aryepiglottoplasty at a tertiary care center

A. Erim Pamuk, Nilda Süslü, R. Önder Günaydın, Gamze Atay, Umut Akyol

Department of Otorhinolaryngology, Hacettepe University Faculty of Medicine, Ankara, Turkey.
E-mail: nildasuslu@yahoo.com

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Laryngomalacia is the most common cause of stridor in neonates and infants. Most cases are mild and resolve spontaneously without treatment. Only severe cases with intolerable symptoms require surgical intervention; in such cases, supraglottoplasty is considered the treatment of choice. The aim of this study was to review and present the outcomes in patients with laryngomalacia who underwent aryepiglottoplasty-a type of supraglottoplasty. The medical records of children diagnosed as laryngomalacia who were followed up at Hacettepe University Hospital, Department of Otorhinolaryngology, between 2007 and 2012 were reviewed retrospectively. The study included 16 children who required surgical intervention. The mean age of the 16 children included in the study was 133 days (range: 7 days-48 months). Among the patients, 9 (56%) were male and 7 (44%) were female. In all, 7 patients (44%) had a comorbid condition. Laryngomalacia diagnoses were as follows: type I: n = 2, 13%; type II: n = 13, 81%; type III: n= 1, 6%. Stridor completely resolved in 10 of the children who underwent aryepiglottoplasty. Three patients required tracheotomy and 3 required revision supraglottoplasty; these six cases were considered as failed surgical treatment. The aryepiglottoplasty success rate was 63%. None of the patients had any intraoperative or postoperative complications. Despite the primarily benign nature of laryngomalacia, comorbid conditions can exacerbate symptoms and negatively affect the prognosis. Aryepiglottoplasty can be performed with high success and low complication rates in properly selected patients.

Key words: laryngomalacia, supraglottoplasty, aryepiglottoplasty.

Laryngomalacia is characterized by inward collapse of laryngeal structures, resulting in a narrow air passage and turbulent airflow during inspiration[1]. Laryngomalacia is the most common cause of stridor in newborns, and affects 45%-75% of all infants with congenital stridor[2]. The typical clinical presentation is high-pitched intermittent inspiratory stridor during the first 2-3 weeks of the neonatal period. The severity of laryngomalacia can be mild, moderate, or severe[3]. Most cases are mild, and present with inspiratory stridor with a coordinated suck-swallow-breathe sequence; such cases do not require therapeutic intervention. Mild laryngomalacia resolves spontaneously within 1-2 years in 70% of patients[4]. Symptoms characteristic of severe cases are poor weight gain, difficultly feeding, choking, post-feeding vomiting, cyanosis (usually exacerbated by agitation and crying), dyspnea with intercostal and suprasternal retractions, and failure to thrive.

Surgical management of laryngomalacia is indicated only in severe cases, and aryepiglottoplasty is the surgical treatment of choice[5]. The aim of the present study was to review and present the outcomes in patients that underwent aryepiglottoplasty at a pediatric tertiary care center and to discuss the management of laryngomalacia.

Material and Methods

The records of patients diagnosed as laryngomalacia at Hacettepe University Hospital, Department of Otorhinolaryngology,

Ankara, Turkey, between 2007 and 2012 were reviewed retrospectively. The study included 16 patients who required surgical treatment and underwent aryepiglottoplasty-a type of supraglottoplasty. Patient records were analyzed in terms of age at the time laryngomalacia was diagnosed, symptoms, disease severity, comorbid conditions, and surgical outcome (Table I).

The severity of laryngomalacia was classified as types I-III, according to a modification of the Holinger classification[1]. This classification is based on conscious transnasal fiberoptic endoscopic observation, as it is more practical before planning the surgery. Type I is inward collapse of the aryepiglottic fold during inspiration, type II is a curled tubular epiglottis, with shortened aryepiglottic folds, which collapses circumferentially during inspiration, and type III is an overhanging epiglottis that collapses posteriorly, obstructing the laryngeal inlet.

The indications for surgery were stridor with respiratory compromise and difficulty feeding with failure to thrive. Pulmonary hypertension, hypoxia, and pectus excavatum were also considered indications for surgery. The supraglottoplasty technique used in this study is based on cutting the short aryepiglottic folds and excising the redundant prolapsing arytenoid mucosa. We performed this surgery (aryepiglottoplasty) via direct laryngoscopy using cold steel. A carbon dioxide laser was not used in any of the patients as we abstained from the possible thermal injury to the surrounding tissues. Aryepiglottoplasty was considered to be successful if post-surgery breathing was normal and no revision surgery or tracheotomy was required during the follow-up.

Results

The mean age of the 9 male (56%) and 7 female (44%) patients at the time of diagnosis was 133 days (range: 7 days-48 months). The primary symptom resulting in presentation for treatment was stridor (100%). In 14 patients (88%), stridor was present at birth, and in 2 patients (12%), it occurred at a mean age of three months. Laryngomalacia diagnoses were as follows: type I: n = 2 (13%), type II: n = 13 (81%), and type III: n = 1 (6%). In all, 12 children (75%) were feeding orally, whereas 4 patients (25%) had significant feeding problems (3 gastrostomy, 1 nasogastric). In

Table I. Patient Characteristics

	Age at diagnosis	Feeding status	Laryngomalacia type	Surgical procedure	Comorbidities	Revision surgery	Postoperative tracheotomy
1	7 days	Oral	Type II	Aryepiglottoplasty	-	-	-
2	7 weeks	Oral	Type II	Aryepiglottoplasty	-	-	-
3	5.5 months	Oral	Type I	Aryepiglottoplasty	West syndrome	+(2 times)	-
4	7 months	Oral	Type II	Aryepiglottoplasty	-	-	-
5	3 months	Oral	Type II	Aryepiglottoplasty	Pierre-Robin syndrome	-	+
6	2.5 months	Oral	Type II	Aryepiglottoplasty	-	-	-
7	8 months	Gastrostomy	Type I	Aryepiglottoplasty	Mobius syndrome	-	-
8	26 days	Oral	Type II	Aryepiglottoplasty	-	+	-
9	2 months	Oral	Type II	Aryepiglottoplasty	-	-	-
10	8 months	Nasogastric	Type II	Aryepiglottoplasty	Cerebral palsy	-	+
11	48 months	Gastrostomy	Type III	Aryepiglottoplasty	Cerebral palsy, epilepsy	-	+
12	1 month	Gastrostomy	Type II	Aryepiglottoplasty	Escobar syndrome	-	-
13	6 months	Oral	Type II	Aryepiglottoplasty	-	+	-
14	3 months	Oral	Type II	Aryepiglottoplasty	-	-	-
15	22 months	Oral	Type II	Aryepiglottoplasty	Cystic fibrosis	-	-
16	14 months	Oral	Type II	Aryepiglottoplasty	-	-	-

total, 7 children (44%) had comorbid diseases, including West syndrome, Mobius syndrome, Escobar syndrome, cystic fibrosis, cerebral palsy, and Pierre-Robin syndrome.

Aryepiglottoplasty was performed at a mean age of 9.6 months (range: 1-48 months). None of the patients had undergone tracheotomy before the surgery. The mean follow-up was eight months. Among the 16 children, 10 (63%) had normal breathing and feeding post-aryepiglottoplasty, whereas 3 (19%) required tracheotomy post-surgery due to ongoing respiratory failure.

The surgical success rate in the patients with comorbid conditions was also analyzed. Surgery was successful in 7 children (44%) without comorbid disease, whereas 3 children (19%) with comorbid disease required tracheotomy post-aryepiglottoplasty. The overall surgical success rate was 63%. None of the patients had intraoperative or postoperative complications.

Discussion

Rilliet and Barthéz[6] (1853) were the first to describe congenital stridor. Sutherland and Lack[7] (1897) published a detailed review of 18 cases that included the first description of congenital laryngeal obstruction. Laryngomalacia, which comes from the Greek word malakia, meaning morbid softening of part of an organ, was first used by Jackson[8] (1942) to describe a disorder in which supraglottic tissue collapses onto the laryngeal airway during inspiration. Laryngomalacia is the most common cause of stridor (45%-75%) in infants and accounts for 50%-75% of all congenital laryngeal anomalies[4,9]. Some theories have been devised to explain the mechanism, but it remains unknown.

Cartilage floppiness due to developmental abnormalities and immaturity, anatomical variation in the neonate larynx, gastroesophageal reflux, poor neuromuscular control, and hypotonia have all been suggested to be etiological in the development of laryngomalacia, but none have been proven to be so. The cartilage immaturity theory has been refuted by Chandra et al.[10] (2001), who reported that the prevalence of laryngomalacia in premature newborns is no higher than that in full-term newborns, and that cartilage is histologically normal in patients with symptomatic laryngomalacia. The anatomical variation theory is based on abnormal localization of excessive flaccid tissue, but in contrast, it has been shown that the same anatomical findings can occur in normal asymptomatic infants[11]. Gastroesophageal reflux has also been considered a factor contributing to laryngomalacia. Several studies reported a high prevalence (50%-100%) of gastroesophageal reflux in laryngomalacia patients[3,12]. It is not clear if reflux is the main cause of laryngomalacia or if laryngomalacia is the result of an alteration in intrathoracic pressure due to the forced effort in breathing. Although severe reflux has a negative effect on the airway, as it causes edema and laryngeal irritation, mild reflux can play an improving role, by decreasing negative intrathoracic intraesophageal pressure[13]. Poor neuromuscular control and hypotonia are other etiological theories for laryngomalacia. Accordingly, laryngeal tone is provided and maintained by the proper functioning of the peripheral sensory afferent reflexes, brainstem, and motor efferent response. This neurological pathway forms the laryngeal adductor reflex, which is a vagal nerve-based reflex that is affected by stimulation of the superior laryngeal nerve. It has been shown that patients with laryngomalacia have impaired sensorimotor function of the larynx due to alteration anywhere in the laryngeal adductor reflex arch[3].

Laryngomalacia often presents with high-pitched, musical, vibrating inspiratory stridor, which first becomes noticeable during the first 2-3 weeks of the neonatal period, peaks at 6-8 weeks, and resolves spontaneously in 1-2 years[4]. Although the cardinal symptom is stridor, there can also be feeding difficulties, choking, post-feeding vomiting, cyanosis (usually exacerbated by agitation and crying), dyspnea with intercostal and suprasternal retractions, and failure to thrive[9,13]. Stridor is usually affected by sleeping position; the prone position reduces symptoms due to extension of the cervical spine, whereas the supine position exacerbates stridor[7,14].

Laryngomalacia was diagnosed via flexible fiberoptic examination in the office or direct laryngoscopy under general anesthesia. Hawkins and Clark[15] performed fiberoptic laryngoscopy 453 times in 264 patients aged ≤4 years and showed the effectiveness of flexible laryngoscopy

in patients with stridor. Botma et al.[16] conducted a study that included 43 infants with symptoms of stridor; laryngomalacia was diagnosed in 35 of the infants, whereas 6 were diagnosed as vocal cord palsy, and 2 were diagnosed as normal laryngeal appearance. These researchers concluded that fiberoptic laryngoscopy can be considered the first-line method for diagnosing laryngomalacia, as it is a cost-effective and sensitive method of examining the larynx dynamically during breathing. In addition, they suggested that direct laryngoscopy and bronchoscopy should be reserved for patients with additional airway abnormalities and patients with failure to thrive. The frequency of these airway abnormalities ranges from 7.5% to 64%[17,18]. In a retrospective study that included 233 children, Mancuso et al.[19] reported that rigid endoscopy is not essential, but Bluestone and Healy[20] noted that fiberoptic laryngoscopy alone is insufficient for diagnosing significant airway lesions. The benefit of using direct laryngoscopy for diagnosing laryngomalacia remains controversial.

Conservative treatment and waiting expectantly are the main treatment options for laryngomalacia due to the nature of the disorder, although severe cases may require surgical intervention. The criteria used to define the severity of laryngomalacia vary by researcher, but the most common indications are stridor with respiratory compromise and feeding difficulties with failure to thrive[4]. Pulmonary hypertension, hypoxia, and pectus excavatum are also considered indications for surgery. The contemporary literature reports that 10%-31% of infants with laryngomalacia require surgical intervention[3,21]. The decision to treat laryngomalacia surgically must be made on an individual basis in consideration of the patient's overall health and development.

Several surgical methods must be considered. Until the early 1980s, tracheotomy and hyomandibulopexy were the only options[22,23], but as technology advanced, endoscopic techniques have increased in popularity. Endoscopic supraglottoplasty using cold instruments was first described by Zalzal[24] in 1987 and subsequently became the most preferred method due to a low complication rate and high success rate, ranging from 80% to 100%[25,26]. In the present study, 10 of 16 patients (63%) that underwent aryepiglottoplasty did not require revision surgery or tracheotomy, and were symptom-free post-surgery. Congenital anomalies and genetic disorders have an incidence of 8%-20%[3,27,28]. Among patients with severe laryngomalacia that requires surgical treatment, 40% are likely to have a comorbid genetic or congenital disorder[3,29]. Comorbid disorders usually have a negative effect on the surgical treatment of laryngomalacia, as seen in the present study¾only 63% of the patients' surgeries were considered successful.

Tracheotomy is an option used to bypass the obstruction usually considered in patients with accompanying neurological or genetic comorbidities. The percentage of patients that require tracheotomy varies; Martin et al.[5] (2005) reported 6.7% and Whymark[30] (2006) reported 14%. In the present study, 3 patients (19%) with comorbid neurological and genetic disorders underwent tracheotomy following aryepiglottoplasty. Although we included 3 patients that underwent tracheotomy after aryepiglottoplasty in the surgery failure group, it is controversial due to the fact that the need for tracheotomy could not be defined clearly because of their comorbid disorders. Surgical failure is defined as the need for revision surgery, failure to thrive, and the need for tracheotomy post-surgery. In the present study, 3 of 16 patients (19%) underwent revision aryepiglottoplasty 3-8 months post-surgery. In the literature, the revision surgery rate ranges from 1.7% (Roger[31], 1995) to 12% (Whymark[30], 2006)·.

Complications associated with aryepiglottoplasty are quite rare, but include bleeding, granuloma formation, postoperative edema and lower respiratory tract infections, supraglottic stenosis, posterior glottic fibrosis, and webs. One study reported that 10 of 136 patients (7.4%) had complications post-aryepiglottoplasty[26]. No intraoperative or postoperative complications occurred in the present study.

In conclusion, laryngomalacia is the most common congenital anomaly of the larynx. It has a benign prognosis, except in cases with comorbidity. Diagnosis of laryngomalacia is based on direct visualization of the larynx via fiberoptic laryngoscopy. Direct laryngoscopy under general anesthesia may also be used.

Candidates for surgery must be chosen in consideration of the severity of symptoms. Aryepiglottoplasty is the primary surgical procedure when surgery is indicated, and it has a high success rate and low complication rate.

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