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LARYNGOLOGY

Laryngotracheal anomalies associated with esophageal atresia: importance of early diagnosis

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Abstract

Objective Esophageal atresia (EA) is the most common congenital esophageal malformation. Airway pathology, in particular, tracheomalacia and laryngotracheal anomalies is a major cause of morbidity and mortalilty in patients with EA. The aim of this study was to report the incidence and type of laryngotracheal anomalies seen in a large series of patients with EA, and to evaluate their impact on the management of children with EA.

Study design Retrospective study.

Materials and methods Retrospective cohort including all patients referred to the EA National Reference Center from January 2002 to December 2014. Airway assessment was based on endoscopy performed before, during and/or after esophageal surgery.

Results One-hundred and fifty-eight patients were included in the study. Endoscopy revealed tracheomalacia in 141 cases (89.2%) and other laryngotracheal anomalies in 43 patients (27.2%). Ninety-six patients (60.7%) presented with persistent respiratory symptoms, including acute life-threatening events in 21 cases, leading to death in 6 cases. A correlation was observed between degree of tracheal collapse and presence of acute life-threatening events. Laryngotracheal surgery was required in 35 cases (22%).

Conclusion Laryngotracheal anomalies are frequently associated with EA and represent an important etiology of morbidity and mortality that can be prevented by early and systematic diagnosis and aggressive management. An early systematic endoscopic evaluation is recommended to coordinate the airway management with the EA surgery.

Keywords Esophageal atresia · Endoscopy · Laryngotracheal anomalies · Laryngeal cleft · Surgery

Abbreviations

ALTM	Associated laryngotracheal malformations
TM	Tracheomalacia
LM	Laryngomalacia
LC	Laryngeal cleft
VCP	Vocal cord paralysis
SGS	Subglottis stenosis
TH	Tracheal hypoplasia
TA	Tracheal atresia
LA	Laryngeal angioma
+	Associated with

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Introduction

Esophageal atresia (EA) is the most common congenital esophageal malformation with an incidence estimated prevalence of 2.4 per 10,000 births [1]. Despite a dramatic improvement of survival during the past 2 decades, airway pathology remains a major cause of morbidity and mortality in patients with EA. Meanwhile, early recognition and a tailored aggressive management may help in preventing complications. While tracheomalacia is a well-recognized

association with EA, very few studies have reported the incidence and characteristics of other associated laryngotracheal anomalies [2–5]. The impact of these anomalies on airway symptoms and airway management remains unclear. The aim of this study was to describe the incidence and type of laryngotracheal anomalies in patients with EA, and to evaluate their impact on the management of children with EA.

Materials and methods

After IRB approval, a retrospective study included all consecutive patients referred to the EA National Reference Center from January 2002 to December 2014, with at least 1 year of follow-up. Study collected information about EA and associated anomalies, morphological airway observations, evolution of respiratory symptoms and the impact of airway anomalies on the management of EA. Morphological airway assessment was based on flexible or rigid laryngotracheal endoscopy performed before, during and/or after esophageal surgery. Flexible laryngotracheoscopy was performed under topical anesthesia in spontaneously breathing patients, and rigid endoscopy was performed under general anesthesia in spontaneously breathing patients. In case of airway malformation suspected during flexible endoscopy, a rigid endoscopy was performed to complete airway assessment.

Clinical evaluation of the airway was performed by an otolaryngologist and a pulmonologist during the systematic annual assessment performed at the national EA reference center.

Results

One-hundred and fifty-eight patients were included in the study (60 girls and 98 boys), with a mean birth term of 37 weeks of GA (range 31–41 weeks). EA was isolated in 68 cases (43%) and associated or syndromic in 90 cases (57%). Demographic data are detailed in Table 1.

Laryngotracheal endoscopy was performed before the esophageal surgery in 79 cases, during the surgical procedure in 48 cases and after the procedure in 31 cases. Endoscopy revealed tracheomalacia in 141 cases (89.2%). The mean degree of tracheal luminal collapse was estimated at 70% (range from 50 to 100%). Associated laryngotracheal anomalies were observed in 43 patients (27.2%) and are reported in the Table 2. Endoscopic assessment also allowed the identification of upper tracheoesophageal fistula in the eight cases with type B and type D EA. Repartition of laryngotracheal anomalies according to EA classification is reported in Fig. 1.

Table 1 Provides demographic data and associated comorbidity

	No. (%)
Gender	
Male	98 (62%)
Female	60 (38%)
Gross classification of EA	
Type A	13 (8.2%)
Type B	3 (1.9%)
Type C	137 (86.7%)
Type D	5 (3.2%)
Comorbidity	90 (57%)
Syndromes	
VACTERL	12
Di George	2
CHARGE	2
Goldenhar	1
Trisomie 21	1
Associated malformations	
Kidney	29
Genitourinary	10
Cardiovascular	22
Vertebral	18
Extremity	4
Gastrointestinal	7
Diaphragmatic hernia	2
Cleft palate	1
Neurological	8
Otologic	6

A postoperative respiratory distress was observed in 33 cases. This was secondary to airway anomalies in 19 cases, pneumothorax in 8 cases, and cardiac malformations in 6 cases. In patients presenting with respiratory distress related to airway anomalies, endoscopic exams revealed tracheomalacia in all cases, subglottic stenosis in five cases, a laryngeal cleft in five cases, tracheal hypoplasia in one case, laryngomalacia in one case, and bilateral vocal cord paralysis in one case.

During follow-up (mean 48 months, range 25–173 months), 96 patients (60.7%) presented persistent respiratory symptoms, including acute life-threatening events (ALTE) in 21 cases (13.3%), leading to death in 6 patients (3.8%). Five of these patients presented with associated cardiac disease, and one presented with tracheal atresia requiring chronic esophageal intubation and ventilation through a distal tracheoesophageal fistula.

In patients presenting with ALTE, a correlation was observed with degree of tracheal collapse (Wilcoxon test, p value = 0.0009) and presence of prematurity (Wilcoxon test, p value = 0.03). Multivariate logistic regression did not demonstrate significant relationship between ALTE and presence

Table 2 Reports type and frequency of associated laryngotracheal malformations

Laryngotracheal malformations	No. (%)
Tracheomalacia	141 (89.2%)
Laryngeal cleft[a]	18 (11.4%)
Grade 1	8
Grade 2	6
Grade 3	3
Grade 4	1
Laryngomalacia	14 (8.9%)
VCP	7 (4.4%)
Unilateral	6
Bilateral	1
Subglottic stenosis[b]	11 (6.9%)
Grade 1	5
Grade 2	4
Grade 3	2
Laryngeal atresia	1 (0.6%)
Tracheal hypoplasia	1 (0.6%)
Tracheal atresia	1 (0.6%)
Crico-pharyngeal angioma	1 (0.6%)
Upper TEF	8 (5.0%)

VCP vocal cord paralysis, *TEF* tracheoesophageal fistula

[a]According to Benjamin's classification

[b]According to Meyer-Cotton's classification

of associated laryngeal, cardiac or neurologic malformations. No significant relation was observed between degree of tracheal collapse and long-term respiratory outcomes.

Corrective airway surgery was required in 35 patients (22%). This included aortopexy in 10 cases, closure of an upper tracheoesophageal fistula in 8 cases, closure of a laryngeal cleft in 11 cases, supraglottoplasty in 4 cases, laryngotracheal reconstruction in 4 cases, slide-tracheoplasty in 1 case, distal tracheoesophageal fistula ligation with proximal tracheoesophageal intubation and ventilation in 1 case, and laryngeal re-innervation in 1 case.

Discussion

Airway anomalies represent a major cause of morbidity during the first postoperative year following esophageal atresia repair. Respiratory problems are a major cause of mortality in the postnatal period and represent the main cause of late death in patients with EA [6].

Moreover, initial respiratory compromise has been described as a predictive factor for high morbidity during the first year of life [7].

Incidence of associated airway anomalies is not clearly identified in the literature as most studies did not distinguish tracheomalacia from other laryngotracheal pathology [2–4]. Our study, the largest study including systematic laryngotracheal assessment, reports a high incidence of laryngotracheal anomalies in patients with EA, which is consistent with previous anatomical and experimental studies [8–10].

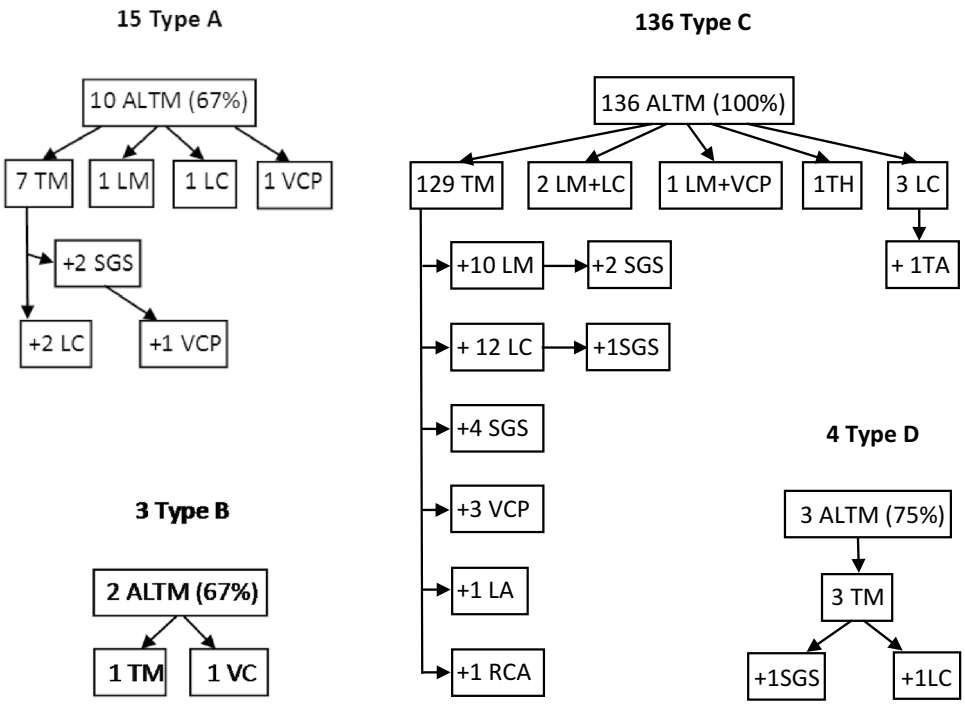

Fig. 1 Repartition of laryngotracheal malformations according to esophageal atresia type. The Fig. 1 reports the repartition of laryngotracheal malformations according to EA type (Gross classification). Excluding tracheomalacia, multiple ALTMs were observed in ten patients

Hseu reported a significant higher incidence of secondary airway malformations, but endoscopic evaluation was performed only in symptomatic patients, possibly explaining this observation [5]. The association of airway anomalies with EA may be explained by a common embryology [11–15].

Tracheomalacia is the most frequent airway anomaly associated with EA, occurring in 62% of patients [16]. Hseu, in a large cohort, reported tracheomalacia in 37.4% of patients, but only 41.1% of patients underwent otolaryngology evaluation [5].

The impact of tracheomalacia is well documented with severe forms occurring in 11–33% of patient with EA and tracheoesophageal fistula [17, 18]. Tracheomalacia requires a surgical correction in 2 to 36.5% of cases [5–7, 19, 20]. The mean age of patients with EA requiring aortopexy was 7 months [21]. The indication for aortopexy was based on the presence of obstructive tracheomalacia on bronchoscopy and the presence of ALTE [21]. In some cases, several aortopexy procedures are necessary to control the bronchotracheal collapse [22]. In patients with tracheomalacia, there was no correlation between their symptoms and the extent of airway collapse. The diagnosis of severe tracheomalacia was made in cases of airway collapse associated with severe airway symptoms and after having treated other possible etiologies of airway compromise such as associated airway or vascular malformations, gastro esophageal reflux, and esophageal stricture.

To date, only few clinical studies focus on airway anomalies associated with EA and their impact on airway morbidity and patient management [5, 23–26]. Airway anomalies such as tracheal atresia, subglottic stenosis, and congenital laryngeal atresia have been sporadically reported [5–7, 27]. Sharma and Srinivas reported bronchoscopic findings in 20.46% of patients with EA including unusual locations of fistulas and laryngeal clefts [26]. Recently, Fraga et al. reported the frequent association of laryngeal cleft and TEF, with or without EA [28].

Symptoms are not specific and life-threatening events may be related to airway anomalies, gastroesophageal reflux, esophageal stricture, and cardiovascular anomalies. A complete assessment of airway anomalies in association with early and aggressive management is necessary to reduce morbidity [29]. Endoscopic examination remains the gold standard for airway assessment, though CT scan may be necessary in cases with vascular anomalies or severe stenosis which cannot be bypassed by the endoscope [16, 30]. To assess airway collapse, flexible bronchoscopy in awake condition may be more accurate than rigid bronchoscopy performed under general anesthesia where airway collapse may be underestimated. Radiological assessment of airway collapse by dynamic CT scanner, as proposed by some authors, appears to be difficult to reproduce and remains challenging to obtain in young children [31, 32]. Moreover, a CT scan may under-diagnose airway anomalies with the absence of fistula localization in 20% of cases. It also exposes children to ionizing radiation [33]. The latter study concluded that CT scans cannot be recommended in the routine assessment of EA.

In our experience, a specific management is required in 20% of patient, often during the perioperative esophageal procedure. This experience confirms the recent results of Sharma's study about the value of systematic laryngo-tracheobronchoscopy in patients with EA to diagnose the associated airway anomalies and to plan management accordingly.

Finally, the goals of pre or peroperative laryngotracheoscopy in patients with EA are:

- To eliminate an associated laryngotracheal malformation to plane the concomitant management.
- To evaluate the tracheomalacia.
- To locate the tracheal fistula and eliminate a second one.
- To place the tracheal tube properly between the fistula and the carina as possible.

Conclusion

Laryngotracheal anomalies are frequently associated with EA and represent an important factor of morbidity and mortality that can be prevented by early and systematic diagnosis as well as aggressive management.

An early diagnosis based on a systematic endoscopic evaluation may be proposed to coordinate the airway management along the EA procedure. We also recommend a long-term respiratory follow-up. When life-threatening events occur, an aggressive management should be proposed, along with addressing-associated anomalies and morbidities.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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