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# Pathophysiology and diagnostic approach to laryngomalacia in infants

S. Ayari[a], G. Aubertin[b], H. Girschig[c], T. Van Den Abbeele[d], M. Mondain[e,*]

[a] Service ORL pédiatrique, hôpital femme-mère-enfant, 59, boulevard Pinel, 69500 Bron, France
[b] Service de pneumo-pédiatrie, hôpital Armand-Trousseau, 26, rue du Docteur-Arnold-Netter, 75012 Paris, France
[c] 173, Route de Desvres, 62280 Saint-Martin-Boulogne, France
[d] Service ORL, hôpital Robert-Debré, 48, boulevard Sérurier, 75935 Paris, France
[e] Service ORL, CHU de Montpellier, 37, avenue du Doyen Gaston-Giraud, 34295 Montpellier cedex 5, France

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**Summary** Laryngomalacia is defined as collapse of supraglottic structures during inspiration. It is the most common laryngeal disease of infancy. Laryngomalacia presents in the form of stridor, a high-pitched, musical, vibrating, multiphase inspiratory noise appearing within the first 10 days of life. Signs of severity are present in 10% of cases: poor weight gain (probably the most contributive element), dyspnoea with permanent and severe intercostal or xyphoid retraction, episodes of respiratory distress, obstructive sleep apnoea, and/or episodes of suffocation while feeding or feeding difficulties. The diagnosis is based on systematic office flexible laryngoscopy to confirm laryngomalacia and exclude other causes of supraglottic obstruction. Rigid endoscopy under general anaesthesia is only performed in the following cases: absence of laryngomalacia on flexible laryngoscopy, presence of laryngomalacia with signs of severity, search for any associated lesions prior to surgery, discrepancy between the severity of symptoms and the appearance on flexible laryngoscopy, and/or atypical symptoms (mostly aspirations). The work-up must be adapted to each child; however, guidelines recommend objective respiratory investigations in infants presenting signs of severity.


Laryngomalacia is defined as collapse of supraglottic structures during inspiration. Clinical practice guidelines for congenital laryngomalacia were published by the SFORL in 2011. This article describes the diagnostic approach to congenital laryngomalacia.

## Pathophysiological mechanisms of laryngomalacia in infants

Not all neonates have the same laryngeal anatomy, but all neonates potentially present supraglottic structures that can invaginate during inspiration. The development of symptoms results from variable combinations of the infant's specific laryngeal anatomy, poor control of the tone of supraglottic structures (either pathological neurological mechanisms or simple physiological variations related to changes in tone during sleep), mucosal oedema, and increased airflow (Fig. 1).

* Corresponding author.
*E-mail address:* m-mondain@chu-montpellier.fr (M. Mondain).

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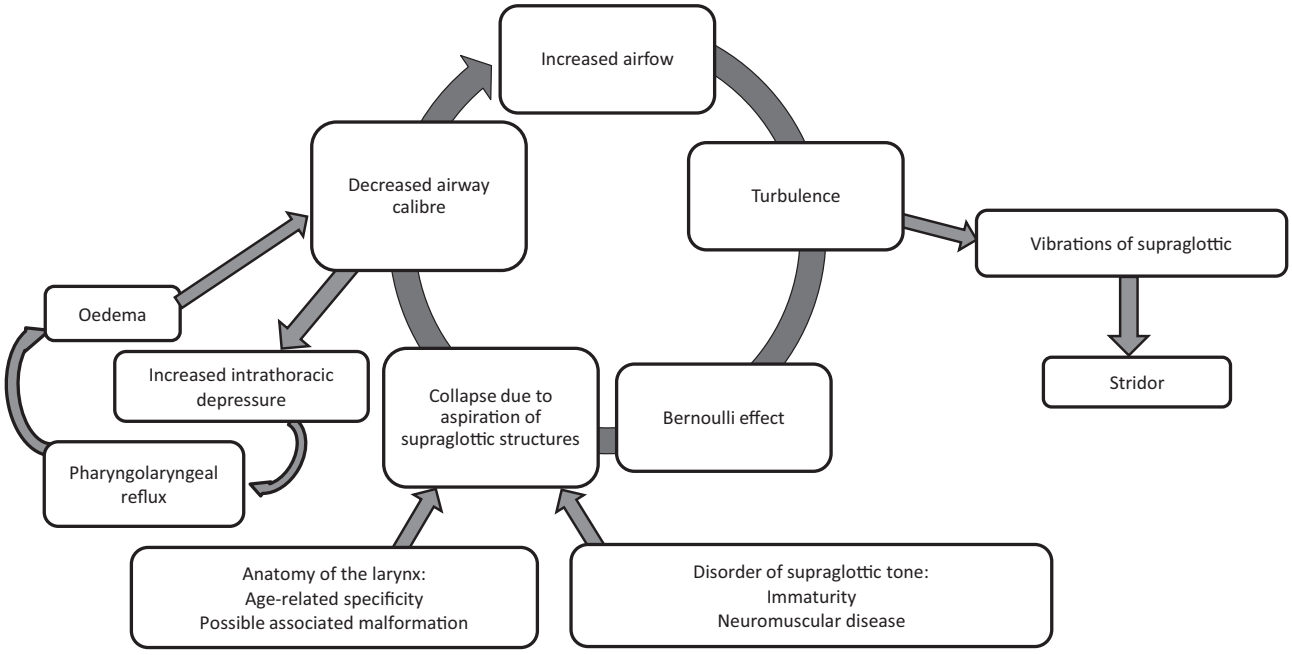


**Figure 1** Pathophysiology of laryngomalacia. $\int$ Larynx anatomy + neurological state + gastro-oesophageal reflux disease (GERD) + Airflow = Symptoms.

The anatomy of the neonatal larynx presents a number of specificities, as the epiglottis is relatively longer (compared to the length of the larynx) than in older children and can be tubular, or even omega-shaped. The infant's epiglottis can therefore prolapse posteriorly and participate in collapse of the supraglottis. Aryepiglottic folds are long, with relatively large, flaccid mucosa. These folds may be short in the anteroposterior plan, drawing the epiglottis posteriorly. Aryepiglottic folds can prolapse medially and inferiorly into the supraglottis, narrowing its lumen. They can also vibrate. Manning et al. showed that the aryepiglottic folds are significantly shorter in infants with severe laryngomalacia than in infants with no signs of severe laryngomalacia [1]. The corniculate cartilages and the superior part of the arytenoid cartilages are clearly visible in the posterior part of the supraglottis, giving the impression of a deep supraglottic interarytenoid groove. These cartilages can prolapse anteriorly and inferiorly into the airway and can also vibrate. During the first 18 months of life, the supraglottis lengthens less rapidly than the rest of the larynx (while the growth of the supraglottic and subglottic airways is identical) [2]: the anatomical structure predisposing to laryngomalacia therefore disappears around the age of 18 to 24 months (Fig. 2).

Mucosal oedema has been demonstrated histologically [3] and participates in narrowing of the airway. This oedema is related either to pharyngolaryngeal reflux (PLR) or mucosal trauma during inspiration. Severe laryngomalacia induces intercostal retraction with increased intrathoracic depression, which, in turn, predisposes to gastro-oesophageal reflux disease (GERD), increasing the mucosa oedema, creating a self-perpetuating process.

The clinical features can be influenced by disorders of neuromuscular tone. Some forms of laryngomalacia are more severely (or exclusively) symptomatic during sleep. The concept of laryngeal immaturity is contested, as laryngomalacia is not more frequent in preterm infants [4] [5]. Documented neuromuscular disease (congenital or acquired with hypotonia and/or psychomotor retardation) can be present with a prevalence varying according to the series between 8 and 50% [6]. The prevalence of neuromuscular disease is higher in the case of severe laryngomalacia and influences the results of surgery [7]. In this setting, laryngomalacia may not be isolated, but part of a broader syndrome of pharyngolaryngomalacia. Moreover, acquired laryngomalacia due to an acquired neurological abnormality (stroke, degenerative disease, tumour) is well known [8].

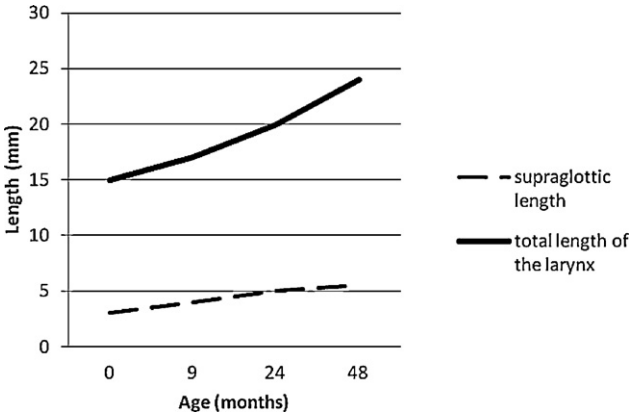


**Figure 2** Course of the length of the supraglottis during growth. Post-mortem anatomical study of four larynges [2] — the supraglottis grows less rapidly than the larynx as a whole.