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UPDATE

Management of laryngomalacia

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Summary Laryngomalacia is the most common laryngeal disease of infancy. It is poorly tolerated in 10% of cases, requiring assessment and management, generally surgical. Surgery often consists of supraglottoplasty, for which a large number of technical variants have been described. This surgery, performed in an appropriate setting, relieves the symptoms in the great majority of cases with low morbidity. However, few data are available concerning the objective results: preoperative and postoperative objective assessment of these infants is therefore necessary whenever possible. Noninvasive ventilation (NIV) may be indicated in some infants with comorbid conditions or failing to respond to surgical management.

Laryngomalacia is defined as collapse of supraglottic structures during inspiration. Most forms of laryngomalacia are minor (70—90%), presenting in the form of isolated and intermittent stridor with no alteration of crying or coughing, no dyspnoea, and no swallowing disorders. These minor forms do not have any consequences on the infant's growth and simply require surveillance by the paediatrician or general practitioner to detect any signs of severity.

Only severe forms of laryngomalacia require therapeutic intervention.

Signs of severity are:

- poor weight gain (probably the most contributive element);
- dyspnoea with permanent and severe intercostal or xyphoid retraction;
- episodes of respiratory distress;
- obstructive sleep apnoea;
- episodes of suffocation while feeding or feeding difficulties.

Endoscopy under general anaesthesia must be systematically performed in these severe forms to confirm the diagnosis and exclude an associated respiratory tract lesion, present in 18.9% of cases in the series published by Mancuso et al. [1]. Associated laryngotracheal lesions (laryngeal dyskinesia, vocal cord paralysis, subglottic stenosis, tracheomalacia) are more frequent in the pres-

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ence of severe laryngomalacia: Dickson et al. [2] reported associated lesions in 79% of cases (with subglottic stenosis in 73.3% of cases and tracheomalacia in 55.3% of cases) in infants with severe laryngomalacia versus 28.8% of infants with laryngomalacia associated with few signs of severity.

Medical treatment and laryngomalacia

Due to the frequency and exacerbating role of pharyngolaryngeal reflux (PLR) in infants with laryngomalacia, anti-reflux treatment should be prescribed, despite the absence of evidence in favour of this approach in the literature (expert opinion).

Lifestyle and dietary measures must always be instituted (thickened milk, maintenance of posture after feeds, no bottle of water before lying down, raising of the head of the bed or mattress), and antacids in infants with regurgitation.

No studies have determined the optimal dose and duration of H2 histamine antagonist or proton pump inhibitor (PPI) therapy or the preferred molecule. Ranitidine can be used at the dose of 3 mg/kg/day and PPIs can be used at the dose of 1 to 2 mg/kg/day. The efficacy of this treatment has been reported in several studies, but no double-blind trials have been conducted [3].

In 2011, the SFORL expert group recommended medical treatment for laryngomalacia with signs of severity, or in the presence of characteristic signs of pharyngolaryngeal reflux on pharyngolaryngeal endoscopy.

PPI therapy is also recommended postoperatively in infants treated by supraglottoplasty procedures: cases of postoperative stenosis have been reported in infants with gastro-oesophageal reflux, but the pathogenic role of gastro-oesophageal reflux simply remains suspected [4].

No study has confirmed the efficacy of local or systemic corticosteroid therapy in laryngomalacia.

Surgical treatment of laryngomalacia

Methods

Apart from tracheotomy, which remained the reference surgical treatment for severe forms of laryngomalacia for many years, several other surgical techniques have been proposed and have been gradually transformed into minimally invasive endoscopic approaches.

Variot [5], in 1898, was the first to propose resection of the excess mucosal tissue on the aryepiglottic folds, based on the post-mortem findings in a neonate with stridor. In 1922, Iglauer [5] was the first to perform partial epiglottectomy in a patient with laryngomalacia with a favourable outcome. In 1928, Hasslinger [5] performed endoscopic forceps division of the aryepiglottic folds in three patients with good results. In 1971, Fearon et al. [6] reported cases of suture of the epiglottis to the base of the tongue, allowing extubation of their patients. During the same period, cases of hyomandibulopexy were reported in France with satisfactory initial results [7], but this technique was subsequently abandoned. In 1981, Templer et al. [8] performed resection of the epiglottis, ventricular folds and aryepiglottic folds via lateral pharyngotomy in an 18-year-old patient with a satisfactory result. The publication by Lane et al. [9] reporting endoscopic treatment of laryngomalacia with resection of excess supra-arytenoid mucosal tissue and the epiglottic mucosa using microinstruments (microforceps and microscissors) led to a renewed interest in these endoscopic treatments. One year later, Seid et al. [10] used the CO_2 laser to divide short aryepiglottic folds in three patients. Following numerous subsequent publications [11—13] endoscopic treatment of laryngomalacia has become the standard treatment.

Anaesthesia

The main methods of ventilation are:

- mechanically controlled ventilation via a small calibre endotracheal tube is rarely used, as it interferes with the surgical procedure;
- spontaneous breathing anaesthesia, which constitutes the technique of choice of experienced anaesthetist-surgeon teams;
- intermittent apnoea technique that provides the surgeon with only a limited time to perform the procedure between two reintubations.

Jet ventilation, sometimes described for this surgery, has not been validated for laryngeal obstructive diseases such as severe laryngomalacia.

Surgical technique

Most of the new surgical techniques used in this disease, called supraglottoplasties, are designed to reduce the excess tissues on supraglottic structures responsible for collapse. Several improvements have been proposed over time, not only concerning the endoscopic surgical technique, but also concerning the methods used to resect excess tissues. Since the first descriptions of supraglottoplasty [9,11—13], technical modifications have mainly concerned the site and extent of the tissues to be resected. The surgical technique [14] usually consists of division of short aryepiglottic folds, and sometimes a resection of excess supra-arytenoid mucosal tissue (Fig. 1), section of the median glossoepiglottic ligament with suspension of the epiglottis to the base of the tongue, partial epiglottectomy [15] or a combination of several of these techniques. Supraglottoplasty is usually bilateral, but some authors have highlighted the advantages and disadvantages of unilateral versus bilateral supraglottoplasty and have reported a lower risk of supraglottic stenosis after unilateral supraglottoplasty [16]. Various methods have also been proposed for resection of excess tissues: cold microinstruments (microscissors), CO_2 laser, Thulium laser [17], diode laser, microdebriders [14] and no differences in terms of the results obtained with these various techniques have been reported in the literature [18]. According to some authors, the advantages of laser and microdebrider compared to microscissors are the absence of intraoperative bleeding, a decreased risk of oedema and simplification of the operative technique. Lasers and microdebriders are more expensive than classical microsurgery instruments and lasers require special precautions to avoid the risk of fire during surgery.