Brachycephalic syndrome (BS) is an established cause of respiratory distress in brachycephalic breeds. Breeds most commonly affected are English and French bulldogs, pugs, and Boston terriers; however, Pekingese, Shih tzu, Cavalier King Charles Spaniels, Boxers, Dogue de Bordeaux, and Bullmastiffs are also categorized as brachycephalic dogs. Most owners report heat, stress and exercise intolerance, snoring, inspiratory dyspnea, and in severe cases, cyanosis and even syncopal episodes. Sleep apneas can be observed, and occasionally gastrointestinal signs such as vomiting and regurgitation.

ANATOMIC AND PATHOPHYSIOLOGIC CHANGES OBSERVED IN BRACHYCEPHALIC BREEDS

Skull Conformation Anomalies

Brachycephalic breeds have a shorter and wider skull compared with mesaticephalic and dolichicephalic breeds, which leads to a compressed nasal passage and additional mucosal hyperplasia and secondary collapse of the upper airway contribute to a multilevel obstruction and the genesis of the so-called brachycephalic syndrome. Surgical treatments usually include widening of stenotic nares as well as various palatoplasty techniques to improve airflow through the rima glottidis. The overall prognosis for a significant improvement is excellent.
altered pharyngeal anatomy. In addition, pugs are reported to have a dorsal rotation of the maxillary bone, miniscule or absent frontal sinuses, a ventral orientation of the olfactorial bulb, and altogether, a shorter craniofacial skull measurement than French and English bulldogs.

This dorsal rotation has been discussed as a potential cause for aberrant nasopharyngeal turbinates, which are also more commonly reported in pugs (Figs. 1 and 2).

**Soft Tissues Changes**

**Stenotic nares**

One typical and easily recognized primary anatomic component of brachycephalic syndrome.

BS-affected dogs have stenotic nares, which reduce each nostril to a vertical slit (Fig. 3).

**Soft palate hyperplasia**

Although the literature used to emphasize an elongated soft palate, fluttering, and obstructing the rima glottidis as a primary component of BS, recent radiographic, computed tomography (CT), and histologic examinations demonstrated an additional pathologic thickening of the soft palate, which might play a major role in the nasopharyngeal obstruction. One study demonstrated a positive correlation between the thickness of the soft palate and the severity of the clinical signs. A recent study using CT evaluation of airway dimension showed a significantly thicker soft palate in French bulldogs compared with pugs but no free airway space dorsal to the soft palate in 81% of pugs. In addition to soft palate hyperplasia, CT and endoscopic studies reported hyperplasia of the nasopharyngeal mucosa, hypertrophy and eversion of the tonsils, and an overlong and thickened tongue (macroglossia), which further displaces the soft palate dorsally.

**Laryngeal, Tracheal, and Bronchial Anomalies**

**Laryngeal diseases**

Laryngeal diseases associated with BS are thought to be mainly secondary to the turbulent airflow and chronic high negative pressures in the pharynx. They include

- Mucosal edema
- Everted laryngeal saccules (ELS)
- Laryngeal collapse

![Fig. 1. Dorsal rotation maxillary bone. Midsagittal CT image of a 4-year-old pug depicting dorsal rotation (arrow) of the maxillary bone.](image)
In one early classification, ELS were considered the first stage of laryngeal collapse\(^3\) (Fig. 4). Stage 2 was characterized by a medial displacement of the cuneiform processes of the arytenoid cartilages, and stage 3 by a collapse of the corniculate processes with loss of the dorsal arch of the rima glottidis. Altogether, the incidence of laryngeal collapse varies from 50%\(^{35,36}\) to as many as 95%\(^{37}\) in BS-affected dogs. Studies report that the size of the rima glottidis is smaller in pugs\(^3\) and that they are also significantly more often affected by severe laryngeal collapse than French bulldogs.\(^3\) In this breed, the arytenoid cartilages can even invert into the laryngeal lumen as a consequence of lack of rigidity (chondromalacia), which makes the larynx incapable to withstand high negative pharyngeal pressures\(^2\) (Video 1).

**Tracheal and bronchial anomalies**

Tracheal hypoplasia,\(^2,18,40\) as defined as a tracheal diameter (TD) to the thoracic inlet (TI) ratio (TD:TI) less than 0.2 in nonbrachycephlic and less than 0.16 in

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**Fig. 2.** Aberrant turbinates. Transverse CT image of a French bulldog depicting aberrant nasopharyngeal turbinates (*arrow*).

**Fig. 3.** Stenotic nares. Stenotic nares in a 2-year-old French bulldog.
brachycephalic dogs, has been described in 13% of BS-affected dogs. The English bulldog has the highest incidence of tracheal hypoplasia among brachycephalic breeds, and tracheal hypoplasia in this breed has been defined as a TD:TI ratio of less than 0.12. Although tracheal hypoplasia increases airway resistance, its contribution to the syndrome is likely minimal.

Bronchial collapse was found to be significantly correlated to the severity of the laryngeal collapse, and pugs were found to be most severely affected. Left-side bronchi were generally more affected by bronchial collapse than the right, with the cranial left bronchus most commonly collapsed. Whether the etiology is loss of rigidity (chondromalacia), increased negative pressure or compression within the chest remains to be investigated (Video 2).

Gastroesophageal Diseases Associated with Brachycephalic Syndrome

Dysphagia, vomiting, and regurgitation are common clinical signs in brachycephalic breeds, and investigation of dogs affected by BS showed concurrent esophageal, gastric, or duodenal anomalies. The negative intrathoracic pressures generated by increased inspiratory effort is believed to be a major cause of gastroesophageal reflux. The associated regurgitation and vomiting can contribute to upper esophageal, pharyngeal, and laryngeal inflammation. French bulldogs exhibit significantly more often and more severe digestive signs than pugs.

DIAGNOSIS

Diagnosis is usually based on owners’ reports, clinical examination, and diagnostic imaging.

Clinical Diagnosis

Snoring, inspiratory dyspnea, cyanosis, and in the most severe cases, syncopal episodes are most often reported by owners. On inspection, stenotic nares and inspiratory efforts with even abdominal breathing can be observed. A particular attention shall be paid to respiratory sounds.

Whereas snoring is most likely caused by air turbulences in the oro-pharyngeal region, the high-pitch sound associated with extreme inspiratory effort is related to more
severe airway compromise when turbulent air is passing through the collapsed larynx or nasopharynx.

**Diagnostic Imaging**

Radiographic, fluoroscopic, CT, and endoscopic studies all contribute to the evaluation of the static and dynamic obstruction of the respiratory tract. In a clinical practice setting, a proper evaluation of BS patients should include at least neck and thoracic radiographs, and endoscopic examination of the upper airways.

- Thoracic radiographs are performed to document secondary heart or lung diseases and to rule out aspiration pneumonia. Also, on occasion a sliding hiatal hernia can incidentally be found on a lateral radiograph.
- Lateral radiographs of the neck (when CT is not available) can help to assess the soft palate thickness as defined by the soft tissue density present between the nasopharynx and oropharynx.
- CT evaluation of the head and neck allows a detailed assessment of the nostrils, vestibule, nasal cavity, and nasopharynx and oropharynx (Fig. 5).
- Endoscopic examination provides more information on the dynamic changes within the upper airways:
  - With the dog intubated, retrograde rhinoscopy performed with a 120° rigid scope or a flexible endoscope allows for good evaluation of nasopharyngeal tissue hyperplasia and collapse as well as for the presence of aberrant turbinates (Fig. 6, Videos 3 and 4).
  - With the dog extubated, a laryngoscopic examination can expose ELS and also help evaluate laryngeal dynamics. With laryngeal collapse, lack of abduction during inspiration or even paradoxical movements of the arytenoid cartilages can occur. In pugs and other dogs affected by laryngeal chondromalacia, the dorsal border of the cuneiform process of the arytenoid cartilages can even invert into the laryngeal lumen (Video 5).

![Fig. 5. Soft palate thickening. Midsagittal CT image of 2-year-old French bulldog with thickening of the soft palate (arrow).](image-url)
CONTROVERSY REGARDING THE GENESIS OF BRACHYCEPHALIC SYNDROME

The genesis of BS is thought to be due to anatomic changes which lead to increased inspiratory resistance. With significant negative pressure, the soft tissues are drawn into the lumen resulting in collapse of the upper airway. Eversion of the laryngeal saccules, nasopharyngeal collapse, and laryngeal collapse, are suspected to contribute to clinical signs and further deterioration of BS, which might ultimately cause syncopal episodes and death from suffocation.

Although, in the past, an overlong soft palate fluttering in the rima glottidis has been considered as the main cause of BS, it remains difficult to estimate the greatest contributor to the clinical signs. The nose is known to be the greatest source of flow resistance in the total airway system, and rhinomanometric studies confirm that intranasal resistance is significantly higher in brachycephalic dogs compared with normal dogs. The major upper airway obstruction was postulated to be intranasal secondary to aberrant turbinates or located in the compressed nasopharynx. But late CT studies comparing pugs and French bulldogs found that the smallest airway space is located dorsal to the soft palate even in dogs with aberrant nasopharyngeal turbinates. Additionally, aberrant turbinates were also described in clinically normal English bulldogs, which means that the contribution of the aberrant turbinates to BS needs to be more thoroughly evaluated.

Although laryngeal collapse has usually been considered to be associated with progression of the disease, a significant correlation between age and severity of laryngeal collapse was demonstrated only recently. In another study including pugs and English and French bulldogs with stage 1 laryngeal collapse, no correlation between Glottic index and age or weight could be demonstrated. Finally, the overall postoperative prognosis of BS was not affected by the grade of laryngeal collapse in 2 recent studies.

Overall, although it is clear that air cannot flow through the nose as long as the nostrils are obstructed, it remains uncertain which part of the obstructed airway—nasal cavity, nasopharynx or rima glottis—is most responsible for the clinical signs associated with BS. In that regard, the effect of soft palate resection might be due to the opening of the nasopharyngeal space and not to the relief of the rima glottidis obstruction.

Fig. 6. Nasopharyngeal turbinates. Images of a retrograde rhinoscopy displaying aberrant nasopharyngeal turbinates.
TREATMENT OF BRACHYCEPHALIC SYNDROME

Medical Therapy

Patients presented with acute signs of respiratory distress should be treated accordingly with cooling, tranquilizers, oxygen therapy, and anti-inflammatory drugs. Whenever digestive signs are observed in dogs with BS, medical treatment including inhibition of hydrogen ion secretion and gastric prokinetic drugs is recommended before and immediately after surgery.

Surgical Therapy

Timing

According to the pathophysiology of the syndrome, relief of the proximally located obstruction should be attempted early to prevent deterioration or possibly reverse tissue collapse, but the optimal time to correct upper airway obstruction has not been determined and was recommended to be performed after the age of 6 months. Recent studies suggest that improvement in clinical signs is still obtained when surgery is performed on mature and middle-aged dogs.

Stenotic Nares

Several surgical options have been described for correction of stenotic nares: amputation of the ala nasi, various alaplasty techniques, alapexy, and vestibuloplasty. Alaplasty is the most used procedure and consists of the excision of a wedge of the ala nasi with primary closure of the defect. This wedge excision can be made vertically, horizontally or laterally. Incisions are made with a No. 11 or 15 scalpel blade or alternatively with a punch. Two to 4 simple interrupted sutures, using absorbable monofilament material, are placed to appose the wedge margins. Hemorrhage resolves quickly when the wound is sutured (Video 6).

Vestibuloplasty has been advocated instead of alaplasty to further improve airflow. It involves the dorsomedial and caudal portion of the ala and results in a wide and open vestibule.

Turbinectomy

Turbinectomy and its laser-assisted variation (LATE) are aimed at removal of malformed obstructive parts of the ventral and medial nasal turbinates. The LATE, combined with vestibuloplasty and staphylectomy, resulted in a decrease of 55% of intranasal resistance 3 to 6 months after surgery compared with preoperative values. Studies show partial regrowth of the removed turbinates but with less mucosal contact points. The long-term positive effects of turbinectomy on intranasal resistance and adverse effects on thermoregulation require further investigation.

Elongated–Hyperplastic Soft Palate

Common surgical techniques for correction of elongated soft palate are aimed at shortening the soft palate by simple resection of its caudal portion (staphylectomy), to prevent it from obstructing the rima glottidis on inspiration. Different landmarks have been recommended, varying from the tip of epiglottis, or the middle to caudal aspect of the palatine tonsils.

During staphylectomy, the caudal border of the soft palate is grasped and held with Allis forceps or stay sutures, and resection of excessive length of soft palate can be performed with a scalpel blade, scissors, monopolar electrocoagulation, carbon dioxide laser, diode laser, or bipolar sealing device (Ligasure, Valleylab, Covidien, Boulder, Colorado).
As these palate-trimming techniques may not address the soft palate hyperplasia, techniques designed to more extensively shorten and thin the soft palate have been described.\textsuperscript{24,73,75,78,79} The folded flap palatoplasty (FFP) has been developed to correct both the excessive length and excessive thickness of the soft palate, therefore relieving also nasopharyngeal obstruction.\textsuperscript{24,78,79} In this technique, the soft palate is made thinner by excision of a portion of its oropharyngeal mucosa and underlying soft tissues. In addition, the palate is made shorter by being folded onto itself until the caudal nasopharyngeal opening is readily visible transorally (Box 1, Figs. 7–9, Video 7).

Postoperative adverse effects or pharyngo-nasal regurgitation have not been observed with the folded flap palatoplasty.\textsuperscript{73,75,79} Whatever technique is chosen, a telescope and high-definition camera system for magnification and illumination of the surgical field (VITOM\textsuperscript{\textregistered}, Karl Storz Endoscopy, Tuttingen, Germany) are helpful (Fig. 10).

**Surgical Treatments for Laryngeal Diseases**

**Everted laryngeal saccules**

Excision of ELS has been described using electrocautery, scissors, tonsil snares or laryngeal biopsy cup forceps.\textsuperscript{1,10,21,32,42,62,69,72} In 1 study reevaluating ELS after single side resection, no regression of the nonremoved site despite treatment of nares and soft palate was found.\textsuperscript{80} Altogether, whether resection of the ELS is needed remains questionable. In several recent BS outcome studies, in which nares and palates were corrected but ELS were either not or rarely addressed, outcomes appeared to be similar to studies in which ELS were excised.\textsuperscript{29,75,79} Also, complications such as laryngeal webbing and regrowth can occur.\textsuperscript{81,82} With this in mind, the authors only recommend removal of ELS when the eversion contributes significantly to the obstruction.

**Laryngeal collapse**

As laryngeal collapse is suspected to be secondary to proximal airway obstruction, first address the proximal obstruction areas (ie, nares and soft palate), as this may obviate the need to treat the collapse.\textsuperscript{2,3,21,23,33,83} In the author’s experience, surgical

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**Box 1**

**The folded flap palatoplasty: surgical technique**

- The head is restrained and the mouth kept open. The tongue is pulled rostrally and maintained with a malleable retractor connected to an articulated arm.
- The caudal edge of the soft palate is grasped with forceps and retracted rostrally and dorsally into the oropharynx, until the caudal opening of the nasopharynx can be visualized.
- The contact point on the ventral mucosa of the soft palate is marked, as this represents the proximal cut of the soft palate.
- The ventral mucosa of the soft palate is then incised in a trapezoidal shape from this mark rostrally to the free edge of the soft palate caudally.
- Laterally, the sides of the trapeze passed just medially to the tonsils.
- The soft tissues under the cut portion of the soft palate are excised together with part of the levator veli palatini muscle.
- The caudal edge of the soft palate is retracted rostrally and is sutured folded on itself with simple interrupted monofilament absorbable sutures.
treatment of laryngeal collapse is considered only when clinical signs do not improve after appropriate treatment of the nares and soft palate. Partial laryngectomy as described earlier has been found to be associated with unacceptably high (50%) mortality rates, and is no longer recommended. Laser-assisted partial arytenoidectomy as recommended for treatment of laryngeal paralysis might provide some relief but needs to be further investigated. Alternatively, arytenoid lateralization is a valid option for dogs with sufficient mineralization of the laryngeal cartilages. On the contrary, its efficacy is questionable in pugs and dogs suffering from chrondromalacia when arytenoid cartilages have a tendency to inwardly rotate during inspiration (pugs). If there is inefficient relief of airway obstruction after the previously mentioned procedures, a permanent tracheostomy can be attempted as a palliative option.

**Removal of other hyperplastic tissues**

Excision of the palatine tonsils has been recommended when they seem to contribute to pharyngeal obstruction. However, the advantage of tonsillectomy warrants further investigation. Similarly, excision of redundant soft tissues located in the pharynx, especially in its dorsal aspect, has been suggested, but more data are needed to evaluate which of the pharyngeal tissues are involved in the obstruction process and the optimal surgical method to remove the hyperplastic tissue.

**Fig. 7.** Folded flap palatoplasty: incision lines in the oral mucosa for the thinning process of soft palate hyperplasia.
Tracheostomy

Although it has been advocated in the past, a preoperative temporary tracheostomy\textsuperscript{1,69,87} is not necessary. Postoperative temporary tracheostomy has been reported in the past in 5\% to 28\% of cases.\textsuperscript{21,29,58,79,88} As the complication rate of temporary tracheostomy in brachycephalic dogs is very high (86\% in 1 study)\textsuperscript{89} it should therefore be reserved for cases not responding to routine postoperative care.

Postoperative Care

The challenge during the postoperative period is to enable adequate airflow in a not yet fully awake patient with potentially swollen airway mucosa. It is critical that BS dogs are monitored constantly after extubation to determine if ventilation is inadequate.

Several methods can be combined or used independently to help relieve upper airway obstruction or improve ventilation after surgical repair:

- The dog can be recovered with the upper jaw hung up, which allows the lower jaw to drop, further opening the airway (Fig. 11).
- Increasing the oxygen delivery—placement of a small nasotracheal tube immediately after surgery but before the dog is awake is a very simple technique of insufflating oxygen, allowing oxygen delivery beyond the rima glottis.\textsuperscript{3,90}
Fig. 9. Folded flap palatoplasty: schematic view showing folding of thinned soft palate upon itself.

Fig. 10. Surgery set-up with telescope. For folded flap palatoplasty, the mouth of the dog is kept open, and the tongue is pushed down using a malleable retractor. Magnification is provided with the Exoscope (VITOM TM, Karl Storz Endoscopy, Tuttlingen, Germany).
It is difficult to gain an accurate perspective of the prognosis for individual dogs afflicted with BS. Most studies evaluating outcome after BS surgery are retrospective in nature, and compare results in different breeds with various combinations of treatments and reconstructive techniques. In addition, these studies compare outcomes with surgical treatments performed at variable patient ages and by different surgeons, using different grading systems. Furthermore, it is difficult to compare the postoperative outcome in these various studies, because there is often a mismatch between the owners’ perception of their pets’ clinical disability and the severity of the clinical signs. A recent study compared preoperative and postoperative treatments in different breeds that underwent the same diagnostic work-up, treatment, and evaluation methods. Despite inherent study limitations, late studies report that around 90% of BS dogs are significantly improved with surgery. This is better than earlier reports. Similarly, perioperative mortality rates have improved from around 15% in earlier reports to less than 4% in more recent studies. Postoperative improvement is most often observed immediately after surgery. Some studies report long-term recurrence of clinical signs in up to 100% of cases, although 89% of dogs still remain improved compared with their preoperative status. In other studies, clinical grades improved in the first 2 weeks after folded flap palatoplasty and remained the same over the following period (mean 12–22 months).
SUMMARY
Animals presenting with BS suffer of multilevel obstruction of the airways and secondary soft tissue collapse. Despite progresses achieved through advanced diagnostic modalities such as CT and endoscopy, the main contributor to the increased inspiratory efforts remains to be found. Recent studies suggest that postoperative prognosis is good even in middle-aged dogs.

SUPPLEMENTARY DATA
Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.cvsrn.2016.02.002.

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