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BRACHYCEPHALIC AIRWAY SYNDROME IN DOGS

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Abstract

Brachycephalic syndrome in dogs, also called congenital obstructive disease of the upper airway is described as a continuous process of anatomical and functional disorders of the respiratory and digestive systems.
The syndrome is characterized by stenotic nares, elongated soft palate, everted laryngeal saccules and in advanced stages by laryngeal collapse.
Clinically, dogs show signs of respiratory distress, such as: severe dyspnea, wheezing, coughing, snoring, exercise intolerance, increased respiratory effort, hyperthermia and collapse, and digestive signs, as: vomiting, regurgitation, ptyalism, pyloric stenosis and inflammation of the gastrointestinal segments.
Medical management includes weight control and reduced physical effort. Patients with acute respiratory syndrome should be treated as an emergency.
Surgical treatment requires wedge resection of stenotic nares or rhinoplasty, shortening of the soft palate or palatoplasty, and removal of laryngeal saccules.
Early recognition and correction of brachycephalic airway syndrome has favorable long-term outcomes for the patient.

Key words: *brachycephalic syndrome, stenotic nares, elongated soft palate, rhinoplasty, palatoplasty.*

INTRODUCTION

Brachycephalic syndrome, also known as brachycephalic airway obstructive syndrome (BAOS), is a well described disorder of the upper respiratory tract of brachycephalic dog breeds (Wetzel J.M, Moses P., 2010). These breeds tend to have respiratory difficulties which are the direct consequence of the anatomical deformities of their head (Roedler F. et al, 2013; Lecoindre R., Richard S., 2004)

Early research on the pathology and treatment of the brachycephalic dogs with respiratory failure were published in the thirties of the last century (Oechtering G. U. et al, 2008).

Primary characteristics of the BAOS include congenital anatomic abnormalities such as stenotic nares, elongated soft palate, hypoplastic trachea and nasopharyngeal turbinates, although, hypoplastic trachea is considered independent of BAOS. Increased resistance during inspiration can cause the development of second changes that include palate and laryngeal edema, swelling, saccule and tonsil eversion, and laryngeal collapse (Meola S., 2013; Trappler M., Moore K, 2011).

All these abnormalities narrow the lumen of the upper respiratory tract and restrict breathing. Often, this ends with asphyxiation and collapse, especially during heat exposure or excitement.

Dogs with more than two of these characteristics present both respiratory and digestive signs (Meola S., 2013; Trappler M., Moore K, 2011).

Diagnosis is usually obvious and uncomplicated, based on clinical signs and physical examination.

Early correction of anatomical abnormalities such as stenotic nares and elongated soft palate may result in a significant improvement of airway dynamics, and may prevent subsequent deterioration during the animal's lifetime (Bray J.,2013).

The purpose of this article is to summarize the current knowledge about BAOS as clear as possible, starting with the genetics, physiopathology, clinical signs and ending with the diagnosis and the medical and surgical treatment.

GENETICS, SEX, AGE AND BREED PREDISPOSITION

No specific genes have been identified. The ancestors of brachycephalic breeds seem to have Asiatic origins.
The brachycephalic dogs have a local chondrodysplasia which is the result of domestication, deliberately kept by the breeders (Koch D. et al, 2003).
The most common brachycephalic breeds are: Chihuahua, Bulldog, King Charles spaniel, Pug, Boston terrier, Maltese, Pekingese, miniature Pinscher, Shih tzu, Yorkshire terrier, and Boxer (Koch D. et al, 2003).
Regarding any possible conection between skull measurements and glottis dimensions in certain breeds, one study made in 2014 demonstrates that in Pug, French Bulldog and English Bulldog no correlation was found. The lack of correlation between skull and glottic indices does not support skull morphology as predictor of glottic morphology. On the contrary, the glottic index was significantly smaller (high and narrow glottis) in Pugs than in English Bulldogs. As a consequence of the lowest glottic index of Pugs when compared to EB and FB, it may be speculated that Pugs original narrow glottis width may predispose to further respiratory deterioration possibly as a consequence of progression to laryngeal collapse (Caccamo R et al.,2014).
Some studies made in 2002 show that there are now sex predisposition , brachycephalic syndrome affecting males and females equally, while some other studies made between 2005 and 2008 show an increased incidence (2:1) in male dogs (Meola S, 2013).
The average age for clinical manifestation of the disease is 2-3 years, although puppies less than 6 months of age have been diagnosticated with severe laryngeal collapse (Meola S, 2013).

ANATOMY

Brachycephalic means “short, wide-headed.” (Evans H, Lahunta A, 2013).
The wings of the nostrils contains fibers of the maxillary labii and nasolabial levator muscles, thing that makes them very maneuverable. Stenotic nares are the result of congenital malformations of the nasal cartilages which cause medial collapse of the alae. This creates a smaller opening at the nostril and a decrease airflow (Koch et al., 2003).
Nasal cavities include 4 meatuses: ventral, dorsal, common and middle, created by the dorsal and ventral nasal choanae and the hard palate. In brachycephalic breeds this are shorten and may contain nasopharyngeal turbinates. This turbinates extend into the nasopharynx, are abnormal and found most in Pugs. Paranasal sinuses commonly miss in brachycephalic dogs (Ginn J. et al., 2008).
The transition from hard palate to soft palate is more caudal to the last molar than in dolichocephalic and mesocephalic dogs. The soft palates is usually elongated and can extend past the epiglottis. This would increase the air resistance at the larynx. The muscular-cartilaginous larynx controls the airflow within the trachea and takes part in vocalization. The narrowest passage of the airflow is the rima glottidis, which is formed dorsally by the paired arytenoid cartilages and ventrally by the paired vocal folds. Laryngeal saccules are located between the vocal and ventricular folds, and normally are not everted (Koch et al., 2003).

PATHOPHYSIOLOGY

All anatomic abnormalities have modified the physiology of the respiratory system of the brachycephalic dogs. Thus, in brachycephalic dogs exhalation is forced rather than passive, because they must overcome the increase in airway resistance. According to Poiseuill's law, a 50% reduction in the radius results in a 16-fold increase in flow resistance (Meola S, 2013).
Stenotic nares (Fig.1) and nasopharyngeal turbinates are considered to be possible stenoses, which lead to an increase in negative pressure during inspiration. They are the most common primary manifestation of BAOS and are found in 17%-77%, respectively 21% of brachycephalic dogs. The negative pressure causes the drawing of the soft tissue into the lumen (stretching of the soft palate), becoming this way hyperlastic, inflamed and the swelling of the airway. Secondary manifestation, such as edema and inflammation of tonsilar tissue, eversion of the laryngeal saccules, partial

collapse of the weakened laryngeal cartilages, narrowed rima glottis, constrict the lumen even more. This way, a vicious cycle develops: collapse of the laryngeal cartilage further decreases of the radius of the airway, which increases the velocity of airflow and the negative pressure in the airway, leading to further collapse (Trappler M, Moore K, 2011;Koch D et al, 2003).

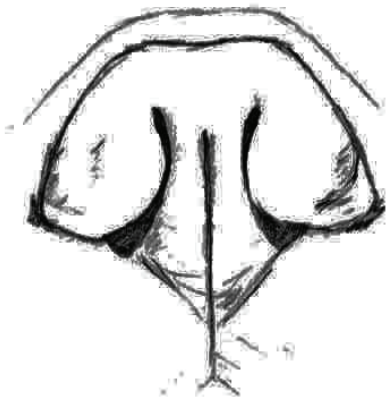

Fig. 1 – Stenotic nares

It is unknown if the elongated soft palate is a primary or secondary event, but it is clearly that the trachea is hypoplastic, due to an abnormal embryogenesis.

Although during inspiration, the soft palate may be caught dorsal to the epiglottis, inciting suffocation, a sympathetically controlled mechanism will cause vasoconstriction, reducing resistance of an acute attack. The wings of the nostrils will be actively dilated, the dog will stretch his neck upward to dilate the nasopharynx and larynx and the air will be inhaled rapidly through the nose, moistened in the conchae, and exhaled though the mouth. Some dogs will have the tendency to open mouth breathe to obtain better airflow, thing that will determine the lateral nasal salivary gland to increase production, to provide enough moisture (Stanley B, 2008).

The respiratory tract is not the only organic structure exposed to increased negative pressure during inspiration. Because of their close vicinity to the airways, the esophagus, auditory canals, central nervous system, and lower respiratory tract should also be examined. Brachycephalic breeds will almost always present an enlarged tongue, difficulty in swallowing, hiatal hernia, gastric bloating, otitis media, neurologic signs, and bronchiectasia (Koch et al., 2003).

In brachycephalic breeds, a possible explanation is the high positive abdominal pressure generated by recurrent vomiting, as well as the negative intrathoracic pressures generated by increased inspiratory work (Dupre G.,2013). Gastroesophageal reflux associated with regurgitation and vomiting can contribute to upper esophageal, pharyngeal, and laryngeal inflammation (White *et al*. 2002). In turn, respiratory distress could stimulate the autonomic sympathetic nervous system, which would slow gastric motility and increase gastric emptying time. Furthermore, the dilated antrum would stimulate gastrin-producing cells responsible for muscular hyperplasia (Peeters 1991; Guilford & Strombeck 1996). The correlation between respiratory and digestive disorders suggests an influence of upper respiratory tract diseases on gastroesophageal diseases, and vice versa. Gastroesophageal disorders (ptyalism, regurgitation, vomiting, and reflux) can aggravate the respiratory signs by encumbering the pharyngeal region and stimulating persistent inflammation. Conversely, chronic respiratory depression promotes gastroesophageal reflux. The close relation between respiratory and digestive problems is sustained by the fact that most of these animals “vomit” large amounts of saliva, when excited or stressed, or during respiratory distress (Dupre G, 2013)

One study made in 2011 concluded that the most important microscopic aspects were hyperplasia and intracellular edema of mucosal lining, diffuse edema, and amplified myxoid matrix in the lamina propria. The anatomy of the palatine musculature of brachycephalic dogs enrolled in the study suffered from extensive degenerative lesions involving the majority of muscular fibers. (Pichetto M et al., 2011)

CLINIC SIGNS

Clinical signs observed in brachycephalic dogs include snoring, inspiratory dyspnea, gagging, productive coughing, difficulty swallowing, sleeping, suffocation, cyanosis and

hyperthermia. Often, they become worse after excitement, exercise, stress or increased environmental temperature. Most of dogs present a reflux esophagitis, regurgitation and vomiting.

DIAGNOSIS

A correct diagnostic can be made only after taking a history, listening of the dog and watching his clinical presentation. Diagnostic imaging as neck and thoracic radiography, CT of the head and endoscopic examination of the upper airways and upper gastrointestinal tract is essential.

Physical examination

Involvement of the soft palate and laryngeal saccules often leads to inspiratory and expiratory dyspnea. Dogs with <50% reduction in airway diameter show an obstructive breathing pattern with slow inspiratory phase and rapid expiratory phase. (Trappler M, 2011)

To perform the examination of the upper airway, it is recommended to anesthetize the animal. This can be done using propofol (3-6 mg/kg) (Trappler M, 2011). The purpose of this is to evaluate the soft palat, tonsils, laryngeal saccules and laryngeal function. In brachycephalic dogs, the soft palate extend past the tip of the epiglottis, blocking the rima glottidis. Everted laryngeal saccules appear as shiny, white, convex structure, cranial to the vocal cords, along the ventrolateral surface of the laryngeal lumen. In pugs, the dorsal border of the cuneiform process of the arytenoids cartilages can invert into the laryngeal lumen. (Trappler M, 2011)

Different studies made along time revealed that the most common manifestation of the BAOS is elongated soft palate.

Radiologic examination

Lateral radiography of the neck can help to assess the soft palate thickness, presented as a soft tissue density between the nasopharynx and oropharynx. Thoracic radiography is made to document secondary heart or lung disease, or, sometimes, a sliding hiatal hernia. (Monnet E, 2013)

Endoscopic examination

Endoscopic examination allows evaluation of the conchaea, soft palate thickness and nasopharyngeal tissue hyperplasia. It is also made to appreciate the stenosis of the vestibulum. In pugs, the dorsal border of the cuneiform process of the arytenoids cartilages can invert into the laryngeal lumen. (Monnet E, 2013)

In 2004, one study made on 30 brachycehalic dogs showed, after performing the endoscopy of the upper digestive airway, that 24 patients presented an hyperplasia of the esophageal mucosa. 13 dogs showed a reflux esophagitis at stage I, 9 at stage II, 2 at stage III and 1 at stage IV compounded by a stenosis of the esophagus, while only 5 of them didn't showed any esophagitis lesions. Sixteen cases of hiatal hernia by sliding or a defective cardiotuberositary position were observed. In 21 cases, the fundic mucosa showed a typical aspect of follicular gastritis. In 13 cases, gastroscopic examination showed an abnormal hyperplasia of the antral mucosa. Five dogs presented no functional or organic abnormalities of the upper digestive tract. (Lecoindre P., Richard S, 2004) (Table 1)

Table 1 – A review based on 4 studies since 2006 to 2010, compared with a review from 2013

Modified parts	Poncet CM, Dupre G, 2006	RecksTW, 2007	Fasanella F. et al, 2010	Wetzell, Moses 2010	Meola S, 2013 Review	Total
Year						2015
Nr. of cases	51	62	90	155		358
Elongates soft palate		54	85	152	62-100 %	81,28%
Stenotic nares	45	36	69	47	17-77 %	55,02%
Everted saccules		36	59	63	53-66 %	44,13%
Laryngeal colaps	14	5		17	53 %	10,05%
Tonsile			50	77	9-56 %	35,47%
Hypoplastic trachea		13		6	13%	5,30 %
Complications	13,3	3,5	10,8	13		11,34%

ANESTHESIA

Before surgery, the patient should be premedicated and stress avoided. Dupré G.,

2013, recommend to performe premedication with Acepromazine (0.01–0.05 mg/kg IM, SC), Dexamethasone (0.1–0.2 mg/kg IM, SC), Opioid analgesic (eg, morphine or methadone, 0.2–0.5 mg/kg IM), Glycopyrrolate (2–10 µg/kg IM), Antiemetics & antacids (SC or IM, depending on drugs) (Dupré G, Findji L., 2013). Some authors recommend the use of dexamethasone in following doses: 0.5-1 mg/kg IV, to avoid the laryngeal edema. Before anesthesia induction, oxygen can be supplied by mask or flow-by for 5-10 minutes. Induction must be swift to allow prompt control of the airway via tracheal intubation. The pharynx should be examined and the larynx assessed for signs of laryngeal collapse.

TREATMENT

The main condition that a brachycephalic dog owner must understand, accept and admit is that this dog breed can not be made normal.

Medical therapy

Medical treatment of a brachycephalic dog include both medical therapy of the upper airways and upper gastrointestinal tract.

Medical therapy of the upper airway is required almost anytime when the patient comes in crises. Hyperthermia should be treated accordingly with cold towels and tranquilizers. It is very important to administrate oxygen therapy, anti-inflammatory drugs (dexamethasone SP 0,22 mg/kg) and to intubate, as the dogs may present glottis edema and cyanosis.

Upper gastrointestinal tract should also be investigated and treated with omeprazole (0,7 mg/kg per os per 24 hours) and prokinetic medication with cisapride (0,2 mg/kg/ 8 hours), immediately after surgery. For severe gastritis and/or duodenitis with parietal fibrosis, the same treatment was advised for 3 months and corticosteroids were added (prednisolone, starting at 0.5 mg/kg per os every 12 hours). (Monnet E., 2013)

Dogs with BAS should avoid activity in warm or humid weather, walks should be kept short and taken at the cool time of day, on a harness to take pressure off the upper respiratory system. (Trappler M, 2011)

No medical therapy will treat the BAOS.

Surgical therapy

Surgery should proceed as soon as possible. An early relief of the proximally located obstruction should be attempted because it is postulated that early correction could prevent, or even reverse more deeply located tissue collapse (Monnet E, 2013).

Stenotic nares

The first procedure for correction of stenotic nares, **amputation of the alae**, was described by Trader in 1949 (Trappler M, Moore K., 2011). It has since been abandoned, because it may be easier than alaplasty technique, but mai result in a lesser opening of the nostril. (Monnet E, 2013)

Alaplasty is the most commonly used technique and it consist of the excision of a wedge of the ala nasi. Incisions are made with a no. 11 scalpel blade and sutured with absorbable monofilament material in 2-4 sutures placed in a simple interrupted pattern.

In the vertical wedge technique (Fig. 2), incisions are started at the apex of the wedge. The medial border of the wedge is parallel to the medial wall of the ala nasi, while the lateral border is made at an angle (40-70^{o}) from the medial border. The opening of the naris will be proportional to the angle chosen. The incision must be deep enough and it must include a portion of the alar fold.

Horizontal wedge resection involves the creation of a wedge in the medial to lateral direction, ending just dorsal to the mucosal edge of the nares. (Trappler M, 2011)

The lateral wedge resection technique (Fig.2) consists of the excision of a vertical wedge of tissue from the caudolateral aspect of the external nose, at the junction between the nose and the skin. The wedge can include a portion of skin or not The wedge is made deep enough to include a portion of the alar fold. When the wound edges are sutured together, the ala nasi is displaced caudolaterally and fixed in an abducted position, thereby opening the nostril. (Monnet E.,2013)

Punch resection alaplasty has been described by Trostel in 2010, in which a dermatologic punch biopsy is used to portion the ala nasi , down to the level of the alar fold. (Monnet E, 2013)

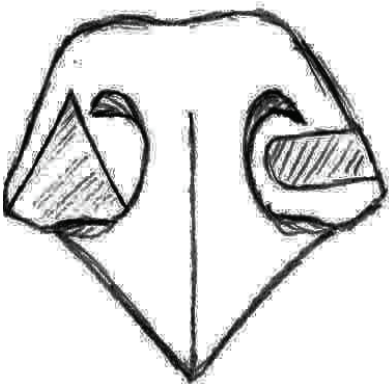

Fig. 2 - Schematic view of the vertical and lateral wedge alaplasty

Alapexy involves removal of the caudal edge of the nares and the adjacent skin (Fig. 3). Two elliptical incisions (5-10 mm long x 3 mm wide x 3 mm deep) are made with a no. 15 scalpel blade, one in the ventral lateral alar and one in the skin 3-5 mm lateral to the ala. The edges of the incision lying closest to each other are apposed with three to four sutures of 4/0 absorbable suture material in a simple interrupted or a simple continuous pattern skin (Fig. 4). The outer aspects of the incision are opposed with three to four sutures of 3/0 or 4/0 nonabsorbable suture material (Fig. 5). (Trappler M., 2011). Alapexy is a good alternative in dogs with excessive flaccidity of the alar cartilage.

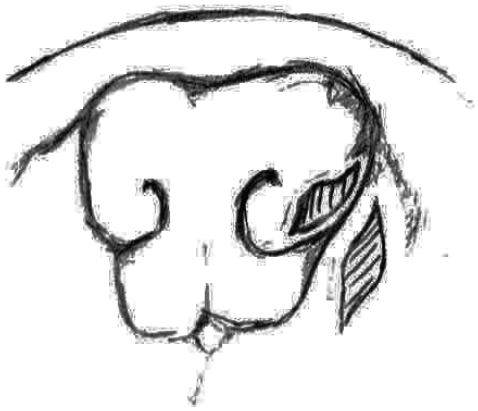

Fig. 3 – The two elliptical incisions made for alapexy

Fig. 4 – The edges of the incisions lying closest to each other in a simple interrupted suture

Fig. 5 – The outer aspects of the incisions

Intranasal obstruction

Turbinectomy is a laser-assisted technique. The aim of this procedure is removal of malformed obstructive parts of the ventral and medial nasal conchae. (Oechtering G. et al., 2008)

Elongated or hyperplastic soft palate

Staphylectomy is the main surgical technique used to treat elongated soft palate and it is described as a simple resection of its caudal portion. The soft palate should be evaluated with the head and tongue in a neutral position and without an endotracheal intubation, because this factors influence the location of the soft palate. If the palate extend past the tip of the epiglottis or the mid to caudal aspect of the tonsillar crypt, then we are talking about an elongated soft palate. When performing the staphylectomy, the caudal border of the soft palate is grasped and held with two stay sutures, placed on either side of the intended line if incision (Fig. 6). It is recommended to perform the section and the sutures in stages (Fig. 7), to avoid complete retraction of the nasopharyngeal mucosa and reduce

hemorrhage. The technique can also done using a bipolar sealing device, a diode laser or a carbon dioxide laser trocautery. Whereas use of a diode laser was found to induce more postoperative edema than a carbon dioxide laser or electrocautery (Dunié-Mérigot *et al*. 2010), no significant differences were found in clinical outcomes when staphylectomy was performed with scissors, carbon dioxide laser, or bipolar sealing device (Davidson *et al*. 2001; Brdecka *et al*. 2007). A very important aspect is that excessive shortening might result in regurgitation during swallowing. It has therefore been recommended to leave the soft palate a bit too long rather than too short (Fig. 8) (Bright & Wheaton 1983; Tobias 2010a,b).

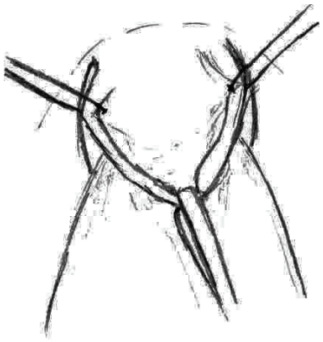

Fig. 6 - The tip of the soft palate is grasped with a forceps and the caudal border is heald with two stay sutures

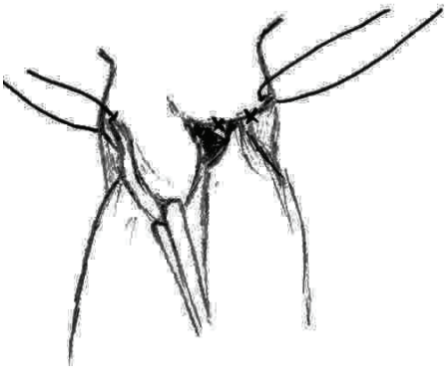

Fig. 7 - The full thickness of the soft palate is incised with a surgical blade approximately half the width of the soft palate.

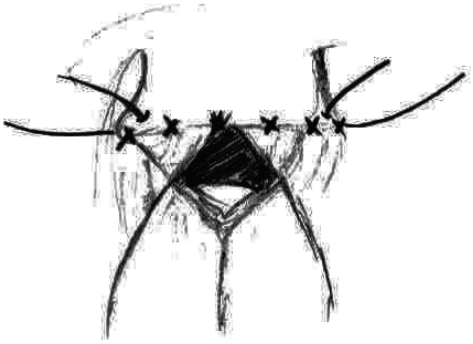

Fig. 8 – The final aspect of the soft palate after staphylectomy

Folded flap palatoplasty technique has been developed to correct both the excessive length and excessive thickness of the soft palate (Findji & Dupré 2008, 2009). In this technique, the soft palate is made thinner by excision of a portion of its oropharyngeal mucosa and soft tissues (Fig. 9) and made shorter by being folded on itself (Fig. 10)(Monnet E., 2013). These techniques leave only a few centimeters of soft palate by using the cranial commissure of the tonsillar crypt as a landmark for simple palatal resection (Brdecka *et al*. 2008; Dunié-Mérigot *et al*. 2010). Postoperative adverse effects or pharyngonasal regurgitation have not been observed with this technique (Monnet E., 2013).

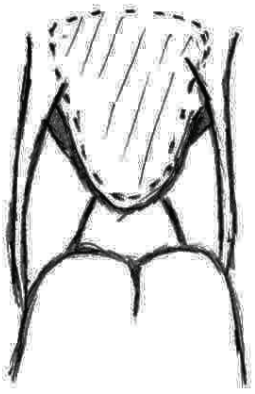

Fig. 9 - Incision lines for the thinning process of soft palate hyperplasia. End of dissection of the soft palate

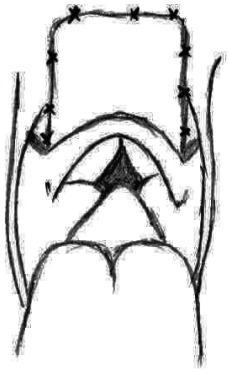

Fig. 10 - Folded flap palatoplasty: after the flap has been sutured

Laryngeal disease

Everted laryngeal saccules are considered the first stage of laryngeal collapse and, generally, are not the most frequent manifestation on BAOS. They are usually excised with a scalpel blade or a scissor. Hemorrhage is usually minimal and can be controlled with gentle pressure, and resection sites are left to heal by second intention. (Riecks TW. Et al, 2007).

Laryngeal collapse is classified into 3 stages; in advanced stages of BAOS (Stages II and III), laryngeal cartilage loses its integrity and collapses inward as a result of the excessive negative pressure created when affected dogs inspire:

• **Stage I** refers to eversion of the laryngeal saccules.

• **Stage II** indicates that the cuneiform processes have come into apposition.

• **Stage III** designation means the corniculate processes are apposed and changes associated with Stages I and II are present. (Lodato D., Mauterer J.. 2014)

It is important to make a difference between laryngeal collapse and laryngeal paralysis.

Tonsillectomy is recommended when they seem to contribute to the pharyngeal obstruction. Because the major palatine artery is present inside the caudal aspect of the tonsillar crypt, 1 ligature of either 4-0 Monocryl or Vicryl can be placed at the caudal edge of the crypt. (Lodato D., Mauterer J.. 2014)

Laryngeal collapse

Stage II laryngeal collapse is defined as loss of rigidity and medial displacement of the cuneiform processes of the arytenoids cartilage. Stage III involves collapse of the corniculate processes of the arytenoid cartilage and loss of the dorsal arc of the rima glottidis. Arytenoids lateralization and permanent tracheostomy are the recommended procedures for severe laryngeal collapse (Trappler M, 2011).

POSTOPERATIVE CARE

The patient will stay intubated as long as he will tolerate the endotracheal tube during recovery. He should be monitored 12-24 hours postoperatively. After 12 hours he can get water in small amount (Lodato D., Mauterer J.. 2014). The diet should consist only soft food for 10-14 days to minimize irritation of the upper airway. (Trappler M., 2011).

Pain management include analgesia, with butorphanol (0,22-0,33 mg/kg iv) and maropitant, for nausea and regurgitation. (Lodato D., Mauterer J.. 2014)

COMPLICATIONS

The most commonly complications are:
Inflammation that can obstruct the larynx and trachea, leading to respiratory distress;
Hemorrhage from the lateral aspect of the soft palate or caudal aspect of the tonsillar crypt;
Coughing and/or gagging;
Nasal discharge;
Voice change;
Regurgitation and/or vomiting;
Noncardiogenic pulmonary edema;
Aspiration pneumonia;
Infection;
Dehiscence (Lodato D., Mauterer J.. 2014)

It is clear that most dogs suffering from brachycephalic syndrome benefit from surgery, according to most recent studies. The proportion of dogs significantly improved by surgery is in excess of 90% (Poncet *et al*. 2006; Riecks *et al*. 2007; Findji & Dupré 2008; Dunié-Mérigot *et al*. 2010; Monnet E., 2014).

CONCLUSIONS

To some degree BAOS is a common finding in most brachycephalic dogs. Early intervention, even as young as 3-4 months of age, should be considered to decrease progression of the

disease and life-threatening laryngeal collapse. (Meola S., 2013). Prognosis depends on the severity of the condition at the time of surgery. Partial resection of the soft palate, laryngeal saccules and nares is expected to relieve moderate to severe signs of respiratory distress in patients who do not have laryngeal collapse (Bojrab M., 2014). Treatment of concurrent GI disease may improve outcomes following corrective BAOS surgery (Trappler M., Moore K., 2011). Emergency management should focus on oxygenation, ventilation, and temperature management for initial stabilization before surgical intervention (Meola S., 2013)

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