BRACHCEPHALIC OBSTRUCTIVE AIRWAY SYNDROME: DIAGNOSIS, MANAGEMENT AND BREEDING CONTROL

Jerold S Bell DVM
Dept. of Clinical Sciences, Tufts Cummings School of Veterinary Medicine, N. Grafton, MA

Short-muzzled breeds have a long history of favor with the public as relatively healthy breeds. However, gradual breeder selection to more extreme brachycephaly has crossed a tipping point predisposing individuals of these breeds to disease predisposition for the brachycephalic obstructive airway syndrome (BOAS).

BOAS is a disorder of breathing difficulty in short-snouted and "bully" breeds, especially the popular (English) Bulldogs, French Bulldogs and Pugs. Other susceptible breeds include Pekingese, Boston Terrier, Shih Tzu, Japanese Chin, and Brussels Griffon. BOAS can cause dyspnea, exercise intolerance, heat intolerance, abnormal and increased respiratory noise, sleep disorders, cyanosis, syncope and death. Clinical signs increase in affected dogs with increasing age. In Persian and Himalayan cats, BOAS causes chronic sinusitis. Individuals in these dog and cat breeds can also have issues with skin fold dermatitis, corneal ulceration, globe proptosis, dental malocclusion, and dystocia.

Of the anatomical components of BOAS, stenotic nares, tortuous sinuses and hypoplastic trachea are inherited. The length of the soft palate and therefore soft palate/epiglottis overlap is also inherited. Soft palate thickness (which exacerbates BOAS) is caused by hyperplasia secondary to chronic turbulence, as are everted laryngeal saccules. Laryngeal collapse and hiatal hernia (with regurgitation) are secondary to the effects of chronic negative inspiratory pressure from restricted airflow.

Compounding the magnitude of BOAS in practice is the popularity of affected breeds. In the past 20 years French Bulldogs went from the 71st ranked AKC breed to the 4th most populous breed, and Bulldogs went from 21st to 5th. Increased public demand produces a greater number of breeders that vary in their commitment to practicing health-conscious breeding. With a rapid rise of breed popularity and BOAS usually presenting in middle age, we will continue to see an increased presentation of BOAS in our clinics.

An issue with BOAS is the "normalization" of brachycephalic stridor to owners and even veterinarians. Some owners will comment that they always know what room their dog is in because they can hear them breathing. However, individual dogs in breeds with extreme brachycephaly die younger (median longevity 8.6 years) than dogs in moderate and non-brachycephalic breeds (median 12.7 years). A higher proportion of deaths in extreme brachycephalic breeds are due to upper respiratory disease (16.7%). Many practitioners have experienced the agonizing presentation of a hyperthermic, cyanotic Bulldog with foamy pulmonary edema preventing the effective delivery of oxygen into their lungs.

ANATOMICAL PRESENTATION AND SURGICAL MANAGEMENT OF BOAS

There are within and between breed variations on the anatomical aspects contributing to BOAS. Stenotic nares is the most frequent observable component of BOAS across breeds, however it occurs at different frequencies. Studies show 75.4% of French Bulldogs have moderately to severely stenotic nostrils, while prevalence amongst Pugs (65.3%) and Bulldogs

(44.2%) is lower. A rule of thumb is that normal combined nares width should be at least 1/3 the width of the nose.

Tracheal hypoplasia is primarily an issue in Bulldogs with some references citing up to 55% of the breed testing affected. On a lateral chest radiograph, the ratio of the tracheal lumen diameter at the thoracic inlet to the width of the proximal third rib should be 2.0 or greater when measured at one year of age or older. The OFA offers a tracheal hypoplasia assessment and database. The frequency of heritable soft palate changes is difficult to assess when confounded with secondary hyperplastic changes.

Three surgical procedures are customarily utilized to improve respiratory function in BOAS affected dogs. Surgical removal of the alar folds of the nares opens up the nostril area. Surgical shortening of an elongated soft palate and resection of everted laryngeal saccules open up the laryngeal airway. A combination of these three procedures based on individual clinical presentation is associated with a favorable long-term outcome in 94.2% of dogs across BOAS affected breeds (though post-surgical complications can be high with inexperienced surgeons).

Across all breeds, the presence of individual anatomical aspects of BOAS do not directly correlate to the clinical presentation of the syndrome; meaning that various combinations of the above, along with non-inherited factors cause a dog to be clinically BOAS affected. Obesity (body condition score) is the most significant non-inherited factor causing clinical BOAS across all breeds. Ambient temperature and humidity also affect the clinical presentation of BOAS, with the majority of emergency veterinary presentations occurring during warmer weather.

PHENOTYPIC ASSESSMENT OF BOAS

With variation in the anatomical presentation of BOAS, research has looked into the diagnostic criteria that separates BOAS affected (BOAS+) and unaffected (BOAS-) dogs. A group at the University of Cambridge, UK led by Dr. David Sargan has developed a closed chamber for Whole Body Barometric Plethysmography (WBBP). A dog's airflow is measured at rest and after controlled activity. Physiological measurements of airflow volume and pattern consistently separate BOAS+ and BOAS- dogs and breeds. Based on WBBP, approximately 50% of study dogs in the three extreme brachycephalic breeds were BOAS affected.

While WBBP is not practical for widespread screening, its results have allowed standardization of other measurements that differentiate between BOAS+ and BOAS- dogs. Standardized working (walking and trotting) tests have been developed by several European groups as respiratory function tests. Most involve standardized, trained veterinary evaluators of breathing function; including a stethoscope examination and body temperature measurements at rest and after exercise.

The British Kennel Club system grades dogs as Grade 0 (normal), and Grades 1-3 for abnormal. Dogs should be over 1 year of age, and should be reassessed every 2 years. Respiratory noise is assessed over the lateral larynx. Exercise consists of 3 minutes of walking at 4 to 5 miles per hour and an immediate post-exercise assessment. Grade 0 dogs have no respiratory noise before or after exercise. Grade 1 dogs have noise only identified by stethoscope after exercise. Grade 2 dogs have respiratory noise evident without a stethoscope

after exercise. Grade 3 dogs have respiratory noise without a stethoscope and increased respiratory effort after exercise. Dogs with respiratory noise at rest are rated Grade 3 and are not given the exercise test. There is considerable between examiner agreement across normal and severe (Grade 3) rated dogs, but some overlap between assessments of Grades 1 and 2. Approximately fifty-percent of dogs assessed are given a BOAS+ grade. It is recommended that dogs rated Grade 3, or those with significant stenotic nares not be used for breeding.

The Finnish and Swedish Kennel Clubs utilize a 1000 meter walking test that must be completed within 11 minutes. Heart rate, respiratory rate and body temperature are assessed before walking, and after a 15 minute “cool-down” period post-walking. All three measurements should return to pre-walking levels within 15 minutes. Their testing also includes a stethoscopic grading system similar to the British, as well as stenotic nares assessment. The French Kennel Club has a 500 meter walking test that must be completed in 6 minutes. For Pekinese they must complete a 250 meter walk in 10 minutes.

Studies show Bulldogs with more severe BOAS walk a shorter distance, more slowly and their recovery from exercise takes longer than those with only mild signs of BOAS. Increases in body temperature during exercise are significantly higher in Bulldogs than in controls. Ambient temperature and humidity during exercise tests tend to affect results in Grade 1 and 2 rated dogs, but not in Grade 0 or 3 rated dogs.

Continuing research and refinement of phenotypic screening of BOAS will assist with pre-breeding assessment of dogs. Some of these test results could in the future be combined into an estimated breeding value (EBV) to compare between prospective breeding dogs.

GENES RELATED TO BOAS

There are several studies into the genes causing BOAS. Groups of BOAS+ and BOAS- dogs can be compared through DNA analysis. Results show that many genes are involved in BOAS making it a polygenic disorder. Therefore the task is to identify if there are single genes that have a major effect on BOAS, or if a panel of genes provides a major difference in the liability to develop clinical BOAS. Such a panel would provide a genomic breeding value (GBV) to compare prospective breeding dogs, and possibly differentiate those with a genetic predisposition to become mildly, moderately, or severely affected with BOAS.

Several genetic studies have been conducted to identify genes associated with skull morphology, brachycephaly and BOAS. Identified genes include IGF1, THSB2, SMOC2, FGF4 and BMP3. However many of these genes are fixed (non-variable) in brachycephalic and BOAS liable breeds. Therefore they do not cause a genetic difference between BOAS+ and BOAS- dogs.

Studies from the Cambridge group into a regulatory gene affecting SMOC2 expression show that it affects the facial skeleton in a dose-dependent manner and accounts for 36% of facial length differences. This group has identified a panel of 11-13 genetic markers that account for 35% of the phenotypic variation in Pugs, 47% in French Bulldogs, and 51% in English Bulldogs. Based on this panel they have been able to predict the most severely affected BOAS dogs in each breed. They also find that those less likely to develop BOAS through GBVs have breed-appropriate longer muzzles and wider nares openings.

CONCLUSIONS

BOAS limits the amount of air that a dog can inhale and in severely affected dogs can cause death by asphyxiation. In others it can cause lifelong labored breathing. While several breeds have a high incidence of BOAS, there is still considerable within-breed variation to enable breeders to breed away from genetically susceptible dogs. Respiratory function grading is becoming the standard screening test for BOAS. The US Bulldog, French Bulldog and Pug parent breed clubs have all requested the OFA to investigate a standardized exercise BOAS assessment test with trained veterinary evaluators. Breeders can incorporate nares assessments and trachea measurements, and select for a breed-appropriate but longer muzzle. Based on research models, genetic testing panels against BOAS liability may be possible.

Part of changing the culture that has caused the rapid popularity and breeding of extreme brachycephalic breeds is to remove the social media fixation on them. In some studies, more than half of all advertising that includes a dog has a Pug, Bulldog, or French Bulldog. The British Veterinary Association has called for a moratorium on advertisements containing extreme brachycephalic breeds.

Dog show judges' education is important to select against the breed extremes of short muzzles and tight nares and to reward moderation of head anatomy. Veterinarians should educate breeders and owners on the morbidity of BOAS. Breeders should use genetic screening in breeding schemes. As environmental aspects influence approximately 50% of the clinical presentation of BOAS, owners can improve their dogs' health by keeping them slim and fit.

The issue of BOAS has developed over time. Breeders did not purposefully select for dogs with impaired breathing. Research shows that within-breed genetic variation exists and there are objective means to improve the genetic health of involved breeds through selection. Breeding bans will not alter the public affection for short-muzzled breeds. Prospective owners should seek out health-conscious breeders that perform pre-breeding genetic health assessments of breeding stock. This will reduce the frequency of BOAS as well as other hereditary diseases and allow healthier dogs and cats.

SELECTED REFERENCES

Liu NC, Troconis EL, Kalmar L, et. al. Conformational risk factors of brachycephalic obstructive airway syndrome (BOAS) in pugs, French bulldogs, and bulldogs. PLoS One. 2017 Aug 1;12(8):e0181928. doi: 10.1371/journal.pone.0181928.

Packer RM, Tivers MS. Strategies for the management and prevention of conformation-related respiratory disorders in brachycephalic dogs. Vet Med (Auckl). 2015 Jun 4;6:219-232. doi: 10.2147/VMRR.S60475.

Riggs J, Liu NC, Sutton DR, et. al. Validation of exercise testing and laryngeal auscultation for grading brachycephalic obstructive airway syndrome in pugs, French bulldogs, and English bulldogs by using whole-body barometric plethysmography. Vet Surg. 2019 May;48(4):488-496. doi: 10.1111/vsu.13159.