Guidelines for the echocardiographic studies of suspected subaortic and pulmonic stenosis

C. Bussadori, C. Amberger, G. Le Bobinnec, C.W. Lombard*

S U M M A R Y

Introduction - Subaortic stenosis (SAS) and pulmonic stenosis (PS) are, with PDA, the most common canine congenital heart defects in almost all epidemiological studies or frequency tabulations of the various European countries and the United States. As dog breeder's associations of highly affected breeds such as Boxers and Newfoundlands are attempting to screen breeding stock clinically as well as echocardiographically and are trying to eliminate these defects through the exclusion of affected individuals from breeding programs, it behooves the group of veterinary cardiologists and echocardiographers engaged in these screening programs to use a standardized approach to the echocardiographic examination procedure. This should warrant obtaining comparable studies with little interobserver variability due to the data acquisition procedure. Such uniformly acquired studies would also be easier to review by a board of experts, should this become necessary in the future. The recommendations stated below are valid for the examinations of dogs as well as cats. The present article represents the combined professional opinions of Drs. Bussadori, Le Bobinnec, Amberger and Lombard.

Required equipment - Any commercial echocardiographic unit with 2.5 to 7.5 MHz sector transducers is suitable for imaging. Transducers with small "footprints" (contact surface) are easier to couple to the chest wall in small patients with narrow intercostal spaces. A so-called cardiac package is also required and provides the software for standardized M-mode, 2D- and Doppler measurements. An ECG-tracing must be included on all recordings. It is mandatory to record clear and artifact-free imaging sequences of adequate length of each required view and of each Doppler measurement on a videotape or digital archive system, with proper patient identification, for later retrieval and documentation.

For the Doppler measurements, *pulsed wave (PW) and continuous wave (CW) capabilities* are required. Color flow Doppler (CFD) is a useful option and allows the rapid documentation and localization of turbulent blood flow, thereby facilitating the placement of the PWD- and CWD- cursors for spectral display and peak velocity measurements. It is however a costly option and not essential for the documentation of neither SAS or PS.

Sedation - The authors highly recommend against tranquilization of the patients, as blood flow velocities are affected by most sedatives. Even if the blood pressure measured under sedation is in the normal range, this does not warrant truly non-influenced blood flow velocity values.

Subaortic stenosis

Echocardiographic anatomy of the left ventricular outflow tract (LVOT)

Exact knowledge of the cardiac anatomy, especially of the LVOT and aortic root area, is essential for making correct patho-anatomical diagnoses for the correlation with any blood flow abnormalities. And, by turning this statement around, an attempt must be made to correlate recognized elevated blood flow velocities anywhere along the left sided outflow tract with patho-morphological particularities of that individual patient.

The left ventricular outflow tract, in its long axis view, can be represented as a truncated cone laid down on its side, with the larger base in the ventricle and the smaller top at the insertion of the aortic valve leaflets, i.e. the semilunar valve ring. The anterior limit (directed towards the right ventricle) is represented by muscular more proximal parts and membranous parts of the interventricular septum. The posterior limit, directed towards the free wall of the left ventricle, is formed by the anterior mitral valve leaflet and higher up its transition into the posterior wall of the aortic root, figure 1.

In a short axis view, the contributing structures depend somewhat on the level of the cross section. In a section close to the semilunar valve ring, the right sided craniolateral wall is formed by the membranous septum and the fibrous trigone of the right coronary cusp, the

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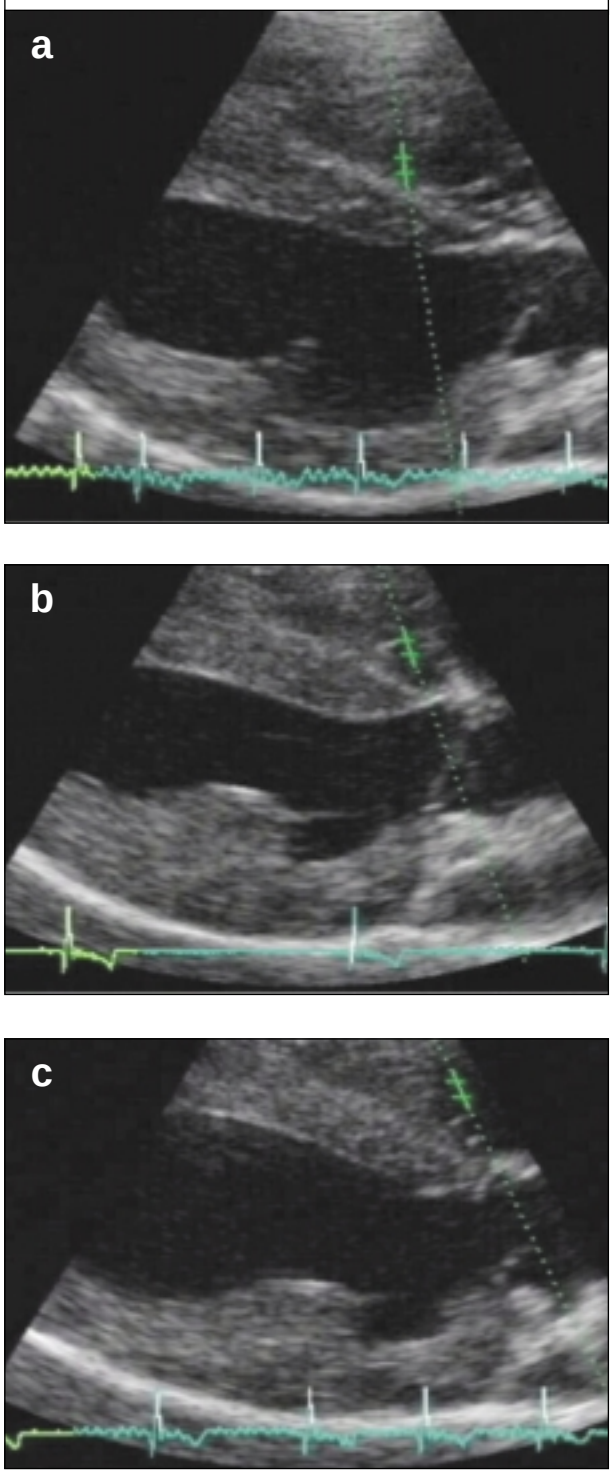
Figure 1 - Long axis image of the left ventricular outflow tract, obtained from a right parasternal transducer placement. The 3 drawn lines illustrate the imaging planes for obtaining the short axis images shown in the following Figure 2.
Figure 1a - Subvalvular.
Figure 1b - Valvular.
Figure 1c - Supravalvular.



cranial limit is formed by the muscular septum, the other, left sided craniolateral wall by the uppermost parts of the left ventricular free wall and the fibrous trigone of the left coronary cusp, and the caudal limit by the anterior mitral valve leaflet. Figure 2 shows several transverse echocardiographic sections of the LVOT of a ca-

Figure 2 - Short axis views of the left ventricular outflow tract, obtained from a right parasternal transducer placement. They were generated by very slight progressive elevation of the imaging plane towards the heart base, as indicated in figure 1.
Figure 2a - Subvalvular, illustrating a circulus fibrous structure forming a "subaortic ring", with a narrower cross-sectional area than the aortic root itself.
Figure 2b - Valvular.
Figure 2c - Supravalvular.

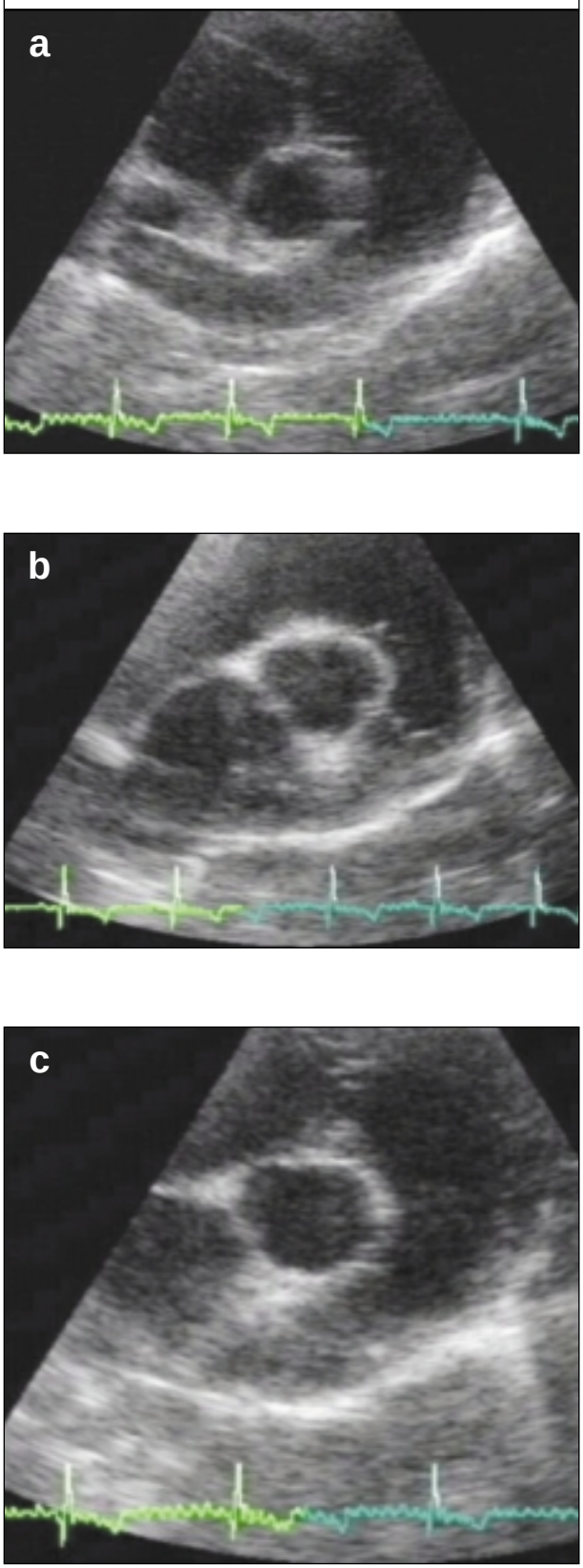
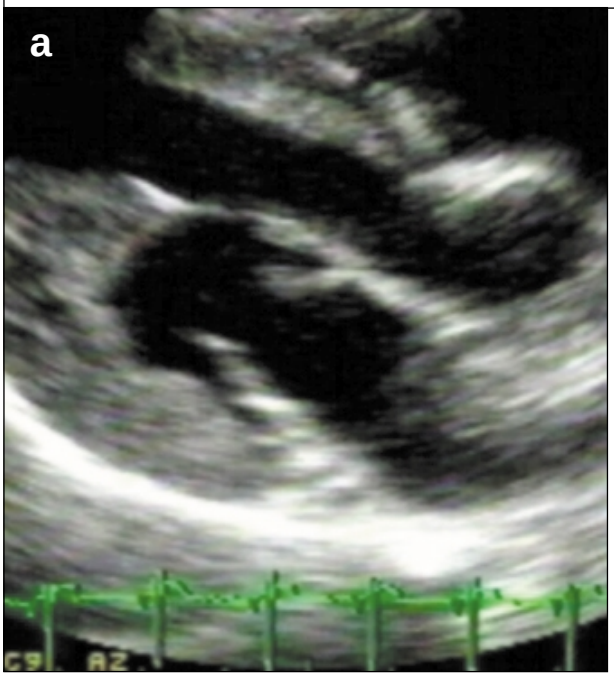
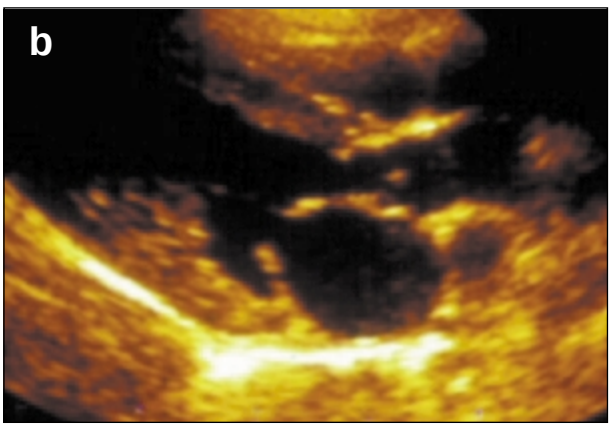
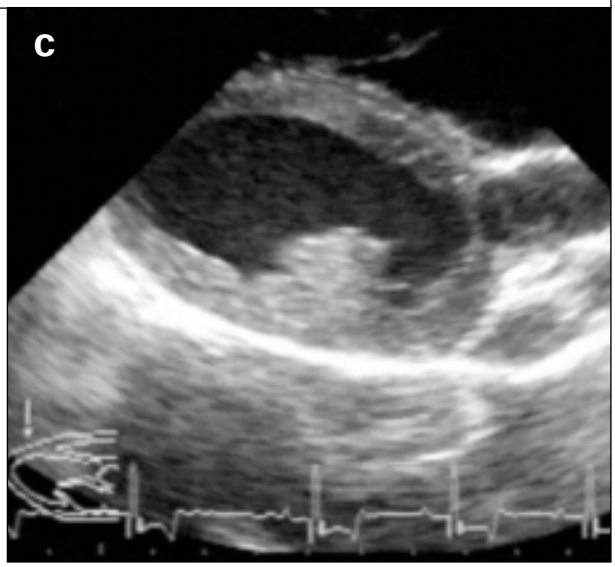
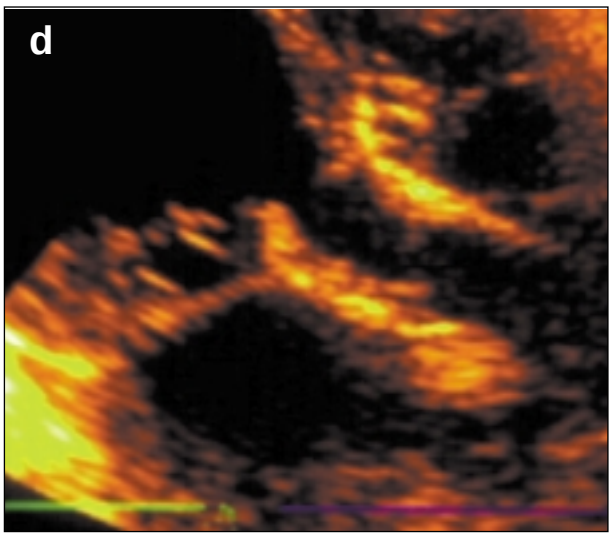


Figure 3 - Long axis image of the left ventricular outflow tract from the right parasternal transducer location.
Figure 3a - "Left ventricular dysplasia" with irregular shape of the LVOT and abnormal chordae tendinae.
Figure 3b - Isolated nodular changes in a Golden retriever with septal thickening.
Figure 3c - Small, echodense fibrous structure forming a subaortic ring below the insertion of the aortic valve in a young Boxer with SAS.
Figure 3d - Tunnel-like formation of a sclerotic obstruction in a Boxer.









nine heart, separated by only a few millimeters elevation of the imaging plane, and optimized for the recognition of the structures contributing to the outflow tract.

Development and presenting forms of SAS

Severe subaortic stenosis with clinical signs of weakness, exercise intolerance and occasional syncope can be found in very young animals of six to eight weeks of age; it is reasonable to evaluate such symptomatic animals as soon as possible. However, the disease more commonly manifest itself and develops its final grade of severity within the first 12 to15 months of life. Therefore, asymptomatic carriers of the lesion may be prescreened at any age, but a definitive assessment of their status for admission to or exclusion from breeding programs should be made only at an age of 12 months and over.

Obstructions occur within the left ventricular outflow tract (LVOT) below the aortic valves (i.e. subaortic stenosis) in over 90% of the cases and are caused by various sized fibrous endocardial nodules or ridges (few millimeters in length) that occur in patches or may form an incomplete or even complete subaortic ring in some animals. In severe subaortic stenosis, the turbulent flow-jet may cause some damage to the aortic valve leaflets themselves, causing them to become thickened and misshaped by the trauma rather than by congenital malformation. Valvular aortic stenosis by fusioned or poorly separated valvular leaflets is rare, and supravalvular aortic stenosis from strictures or coarctation is even rarer. A relatively small aortic diameter without obstructions, representing some degree of aortic hypoplasia and associated with cardiac murmurs and elevated

and turbulent aortic blood flow velocities, appears to occur in the Boxer breed and probably isolated representatives of other breeds as well. In specific breeds (i.e. boxers and retrievers), it's also possible to find a real "left ventricular dysplasia" with an irregular shape of the LVOT and important anomalies of mitral chordae tendinae, some of them can be directly attached to the SAS ring (Fig. 3).

Imaging and Doppler-measurements procedures for SAS

It is mandatory to obtain the standard long and short axis imaging views from the right side as well as the left apical 4 and 5-chamber views, recommended by the echocardiography committee of the specialty of the cardiology of the ACVIM (Thomas et al, 1993). Additional imaging planes may be useful for an optimized anatomic representation of a particular lesion, but are not mandatory. We strongly recommend a complete echocardiographic examination with investigation of both cardiac septum and all four valves, respectively their inflow and outflow tracts, in order to recognize possible coexisting cardiovascular lesions! (SAS can be associated with PS, VSD, MD and PDA). The general guidelines for performing Doppler echocardiography, reviewed by Bonagura et al 1998, are generally accepted and should be followed closely.

Right-sided transducer placement. Ideally, the animal is placed in right lateral recumbency on an echocardiographic table with a cut-out and the transducer coupled to the right thoracic wall from below. Views obtained with the patient standing are acceptable if longer imaging sequences without motion artifacts can be recorded. Clipping of the hair is recommended, but not mandatory, as long as high quality images can be obtained.

The standard right parasternal long and short axis views, optimized for the LVOT, should be recorded. In the long axis view, we recommend to screen the LVOT with color flow Doppler (CFD) for any systolic turbulence that originates in the subaortic area. Such images may help in recognizing discrete lesions mentioned in the paragraph "development and presenting forms" above, see also figure 3. Imaging the proximal parts of the aortic root allows the recognition of the sinuses of Valsalva and often the ostium of the left coronary artery, any valve leaflet motion particularity such as systolic fluttering or midsystolic closure by M-mode imaging, "doming" effect (incomplete opening) in cases with fused leaflets, and any abnormality of the sinotubular junction or any poststenotic dilatation.

In the standard right parasternal short axis views, the LVOT is screened for a progressively narrowing truncated cone in cross-sectional images, see figure 2. Abrupt narrowing, caused by a subaortic fibrous ridge that may form a ring-like structure with a smaller cross-sectional area than the aortic root itself, may be recognized in these imaging planes. Otherwise, the short axis views are more suited to recognize eccentric closure lines of the valve leaflets or any other valvular deformities such as a bicuspid aortic valve, a defect common in man but extremely rare in dogs and cats. In severe forms of SAS, valvular deformities are often "jet lesions" from subaortic turbulences, and not primary anomalies.

From the same short axis views, images and Doppler measurements of the right ventricle, RVOT and pulmonary trunk are recorded, see the section about pulmonic stenosis later in this manuscript. It is important to underline that in the French and Italian survey of SAS, PS is associated in 8% (Le Bobinnec et al.) and 4.4% (Bussadori et al) of all the cases.

Subcostal transducer placement. Subcostal views of the heart can be obtained with the patient in right or left sided recumbency, or even standing. But the authors strongly recommend the right lateral recumbency, as it appears the easiest way, and can follow the right parasternal investigations without repositioning of the patient.

It is relatively easy to obtain a modified, usually 3 to 4 chambered view (left ventricle, parts of the right ventricle or atrium, parts of the left atrium and LVOT with aorta, Fig. 4), and to obtain near perfect parallel alignment of the Doppler cursor with the LVOT. The PWD-window can then be placed just below and sometimes also above the aortic valve plane and allows to obtain good flow tracings and measurements from both locations. For technical reason, the use of PWD is not always possible in the subcostal view. CWD is acceptable, but whether PWD, HPRF or CWD is used, this should be recorded. Figure 6 shows Subcostal Doppler recording of a Boxer with SAS.

Doppler measurement of peak LVOT-velocities from subcostal views must be obtained for suspected subaortic stenosis, as this transducer placement and alignment with the LVOT has shown to consistently provide the highest velocity values in dogs with this malformation (Lehmkuhl and Bonagura 1994). It is advisable to use transducers with 3.5 mHz or lower frequencies to assure sufficient penetration of the tissues and adequate signal strength of the Doppler recordings, as the distance to the aortic valves is quite far in large dogs with this imaging view, see figure 4. The examination is completed with CWD- and CFD-recordings of the blood flow through the LVOT.

Left apical transducer placement. The patient is ideally placed in left lateral recumbency and the transducer again coupled to the left thoracic wall from below for obtaining standard left apical 4- and 5-chamber views (Fig. 5). Alternatively, good quality views obtained from the patient in a standing position are acceptable. From a 4 chamber view, velocity tracings of diastolic flow entering both ventricles (transmitral and transtricuspid flow curves with respective peak velocities) should be obtained and recorded in a pulsed wave Doppler mode, with the Doppler cursor placed just inside the respective ventricle, above the tips of the AV-valve leaflets. Both

Figure 4 - Subcostal imaging of the heart, generating a 3-chamber view (LV, LA and Ao), from a Golden Retriever with SAS. The placement of the PWD-Window in the LVOT just below the aortic valve is illustrated, and a CWD-velocity tracing of more than 5 m/sec is displayed.

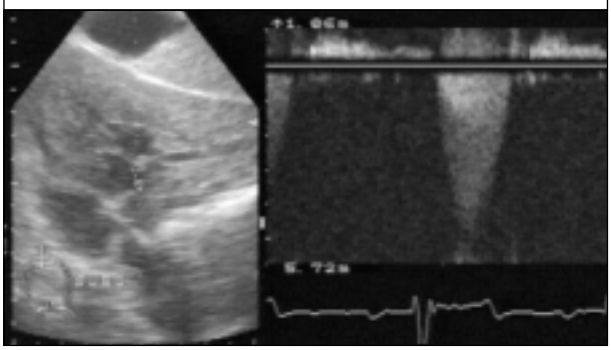
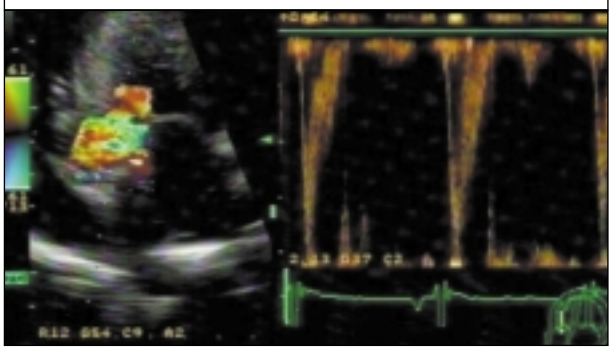


Figure 5 - Left apical 5 chamber view, with the pulsed wave cursor placed just below the aortic valve plane, and the spectral Doppler signal obtained from this location. A clean spectrum is displayed, with a peak velocity of 2 m/sec.



valves must be screened for systolic regurgitation with PWD/CWD and if possible CFD. From a 5-chamber view, the LVOT should be investigated with all 3 forms of Doppler for stenosis and eventual insufficiency. It is always possible to place the PWD-cursor in the LVOT just below the aortic valve plane, and usually also possible inside the aortic root to obtain velocity tracings from both localizations for comparisons.

Measurements obtained from left apical views may be recorded as well for comparative purposes, but should only be accepted if subcostal images and measurements are not obtainable under any conditions.

Normal values for LVOT-blood flow velocities, and classification of recognized lesions.

Several authors have published normal values for LVOT-blood flow velocities in dogs and cats. They are summarized in table 1. Unfortunately, details about the heart rates of the patients, their state of relaxation or excitement, angle of correction for non-perfect cursor alignment with the axis of the LVOT, and a few other potentially important details were not standardized or not indicated in each study. Additionally, there is considerable disagreement among cardiologists about what should be accepted as the upper limit of LVOT-velocity in the canine, and perhaps acceptance of different upper limits in different breeds is necessary. This detail should be settled regionally between the respective groups of cardiology experts working with the breeder's association before starting a screening program.

When elevated peak velocities are found in the LVOT, the values are then transformed into an estimated pressure gradient with the simplified Bernoulli equation. The obtained values are classified as follows:

Mild stenosis:
peak gradients from 20 to 49 mmHg
corresponding to a velocity of 2.25-3.5 m/sec
Moderate stenosis:
peak gradient from 50 to 80 mmHg
corresponding to a velocity of 3.5-4.5 m/sec
Severe stenosis:
peak gradient above 80 mmHg
corresponding to a velocity over 4.5 m/sec

Table 1. Reference values for Doppler-derived LVOT/aortic flow velocities.

Range m/sec	peak +/- SD m/sec	Type	Transducer placement/ sample vol./angle corr.	Nr. of dogs	Sedation	Reference
1.04 -1.38	1.18 +/- 0.11	CW	L	n=20		Yuill & O'Grady
	1.19 +/- 0.18	PW		n=28		Gaber
0.65 - 1.37	1.06 +/- 0.21	PW	L.apical/aorta/no	n=28	YES	Brown et al.
1.06 - 2.29	1.57 +/- 0.33	PW	L apical/aorta/yes	n=50	no	Kirberger et al.
0.73 - 2.01	1.24 +/- 0.30	PW	L.apical/LVOT/yes	n=50	no	Kirberger et al
0.99 - 2.10	1.49 +/- 0.27	CW	L apical/ /yes	n=50	no	Kirberger et al.
< 1.70	1.20	PW				Bonagura & Miller
	1.19 +/- 0.24	n.i.	L or R apical/n.i./n.i	n=20	no	Darke et al
0.92 - 1.50	1.15 +/- 0.15	PW	L apical/aorta/no	n=15		Bonagura et al
1.10 - 2.12	1.62 +/- 0.23	CW	L.apical/aorta/no	N=43 (Boxers)	no	Le Bobinnec 1999 unpub
1.20 - 2.65	1.96 +/- 0.26	CW	Subcostal/…./no	N=72 (Boxers)	no	Bussadori 2000 unpub.

It is important to additionally document and classify any recognized patho-anatomical abnormalities of the LVOT, as well as the morphological abnormalities of the spectral recordings for the differentiation of static and dynamic stenosis. The anatomical classification is based upon previous necropsy and catheter studies from Pyle & Patterson (1984) as follows:

Class 1: (Fig. 3b) thickened and raised septal endocardium to form small 1-2 mm nodules, sometimes also present on the ventricular surfaces of the aortic cusps. This corresponds to a discrete SAS-echocardiographic classification (type 1) with no or only very discrete endocardial thickening (increased echodensity) or rising.

Class 2: (Fig. 3c) Fibrous thickening and elevation of the subaortic endocardium extends from septal portions to the base of the anterior mitral valve leaflet, forming a partial or complete fibrous ring below the aortic valve (echocardiographic classification type 2).

Class 3: (Fig. 3 d) Endocardial thickening and elevation extends circularly all around the LVOT and also in its entire length, forming a concentrically narrowing tunnel and corresponding to an echocardiographic classification type 3.

Pulmonic stenosis

Echocardiographic anatomy of the right ventricular outflow tract (RVOT)

The RVOT can again be represented as a truncated cone, but somewhat curved like the middle parts of a "croissant". The larger base lies in the right ventricle, and the shorter top is formed by the semilunar valve plane. The medial limit (about 30-40% of the circumference, directed towards the left ventricle) is represented by muscular parts of the interventricular septum, and higher up the aortic wall. The lateral limit (about 60-70% of the circumference) is formed by the free RV-wall itself, and above the plane of the semilunar valve by the lateral wall of the pulmonary trunk. In cases of severe PS, considerable lateral bulging of a poststenotic dilatation of the most proximal parts of the pulmonary trunk can be visualized.

Development and presenting forms of PS.

Pulmonic stenosis, in contrast to subaortic stenosis, usually presents with its definite grade of severity early in the life of the affected puppy, because the valvular form of this malformation is much more common. Subpulmonic muscular hypertrophy accompanies the congenital valvular stenosis to some degree, but does not appear to increase in severity during adolescence. The valvular PS can be differentiated into 2 main types:

Type A. (Fig. 8) The annular size is normal. Various degrees of leaflet thickening with incomplete separation of the commissures to almost complete fusion do occur. It causes a systolic doming of the valve ("windsock"-type image) with most often eccentric valvular opening with various degrees of reduced cross-sectional area. Poststenotic dilatation of the pulmonary trunk is invariably present with various degrees of severity. This type is ideally suited for balloon valvuloplasty procedures.

Type B. (Fig. 9) The pulmonary ostium is hypoplastic, with various degrees of valvular leaflet thickening and immobility, but little commissural fusion. The main pulmonary trunk is also often hypoplastic, and rarely has a poststenotic dilatation.

Subvalvular obstructions by a fibrous ring, analogous to SAS, appear to be rare and are invariably associated with additional valvular deformities. Various degrees of fibromuscular infundibular hypertrophy is the more common form of subvalvular PS. Likely, it has a dynamic component and leads to worsening of the stenosis with exercise or during stress tests. In some breeds, especially the English Bulldog and in some Boxers, an anomalous R2A-type left coronary artery (Buchanan 1990) encircles and constricts the right ventricular outflow tract. This anomaly obviously represents an exclusion criterium for any valvuloplasty procedure, as a dilatation of the infundibulum invariably would lead to coronary iscemia and the patients demise.

Isolated supravalvular PS appears to be extremely rare.

Symptomatic animals may be screened at any age. Because the combination of SAS with PS occurs in a considerable number of Boxers (Fig. 6), this breed should be screened for congenital heart defects between 12 and 15 months of age, as proposed above.

Imaging and Doppler-measurement procedures for PS.

The right ventricular outflow tract (RVOT) is nearly impossible to image in its entire length with a perfect long axis view, because of its curved course.

Right-sided view. The animal is ideally placed in right lateral recumbency on an echocardiographic table with a cut-out and the transducer coupled to the right thoracic wall from below, as discussed above. An oblique, somewhat tangential section of the RVOT, obtained from a right parasternal short axis view, can be generated in all dogs. The PWD-window can usually be placed below, and in many cases above the pulmonic valve into the pulmonary trunk. Although this represents only a tangential section of the RVOT, satisfactory Doppler tracings can be recorded from almost all patients with this view, see figure 7. The diameter of the pulmonic annulus should be measured in this view, and a ratio of the diameters of the aortic annulus divided by the pulmonic annulus allows the confirmation of pulmonary hypoplasia (calculation of aortic to pulmonic annulus ratio, see above). Additional Doppler recordings are obtained with CWD and CFD from the same area, as indicated by any suspected or visualized abnormalities.

Figure 6 - SAS and PS from a young Boxer.

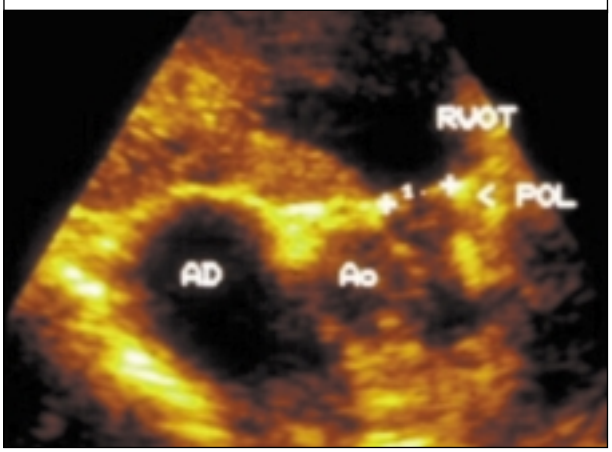
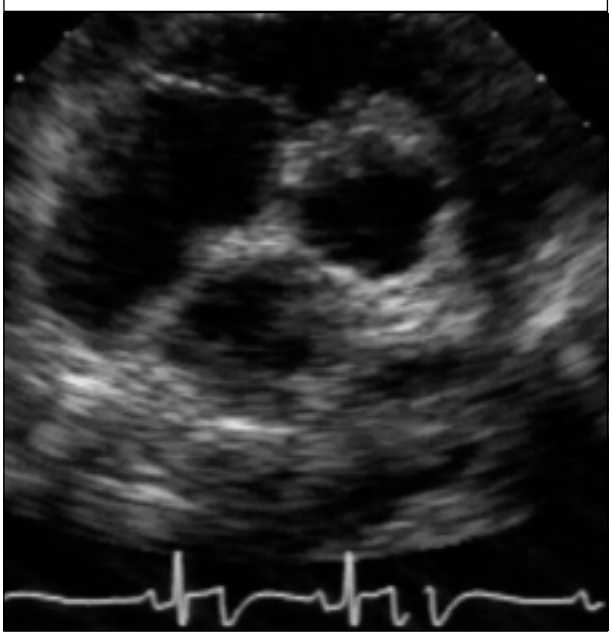


Figure 8 - Right parasternal short axis view from a dog with severe valvular PS. Pulmonic valves are misshaped and distorted. The main pulmonary artery is hypoplastic and shows severe post-stenotic dilatation (Type A).



In patients with subvalvular PS, this view is most often sufficient to image subvalvular narrowing caused by muscular hypertrophy and infundibular as well as valvular malformations. Figure 8 shows valvular malformations with a slight hypoplasia of the main pulmonary trunk and mild poststenotic dilatation. In some cases of valvular PS however, the only visual abnormality is a wide RVOT with a thickened, hypertrophic free RV-wall. The valvular and supravalvular lesions are too proximal to be seen properly. Despite the lack of visualization, the CW-Doppler interrogation reveals an increased maximal outflow velocity, and commonly some pulmonic regurgitation. As an alternative, left sided views may be attempted in such patients.

Left cranial parasternal short axis view. For this view, the patient is placed in left lateral recumbency and the transducer coupled to the left thoracic wall

Figure 7 - Right parasternal short axis view of the RVOT and proximal pulmonary trunk from a dog with pulmonic stenosis. The PW-gate is placed in the RVOT just below the pulmonic valve plane, and CWD-velocity tracing of 6 m/sec is displayed.

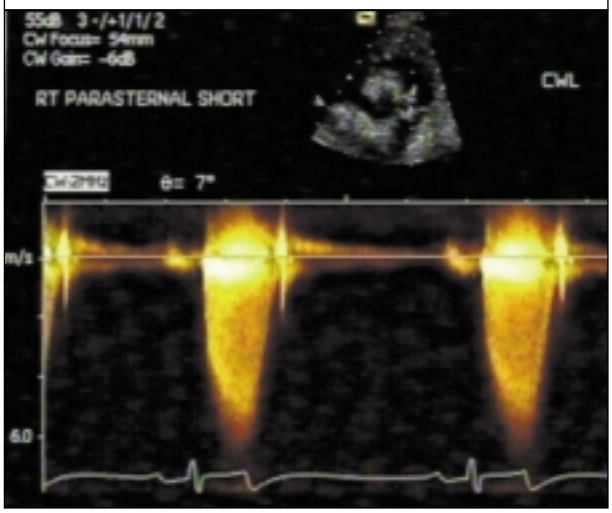
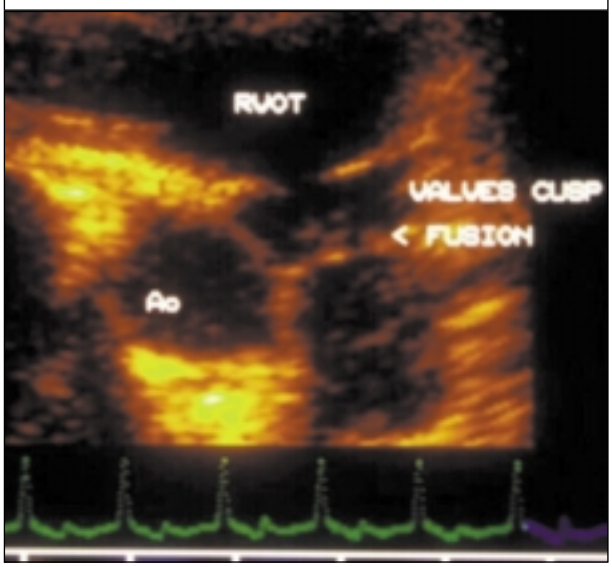


Figure 9 - Right parasternal short axis view from a dog with Type B PS in a young Boxer.



from below, but in a parasternal position, away from the apex. After obtaining a long axis view of the aortic root, a steep dorsal and slightly more cranial angulation often lets the RVOT appear in view with the main pulmonary artery in most of its length and even the bifurcation into left and right main pulmonary arteries. This view displays valvular abnormalities and poststenotic dilatations nicely. The Doppler interrogations are then performed.

Normal values for RVOT-blood flow velocities are presented in table 2. When elevated peak velocities are found in the RVOT, the values are then transformed into an estimated pressure gradient with the simplified Bernoulli equation, the same way as with SAS. The obtained values are classified as follows:

Table 2. Reference values for Doppler-derived RVOT/pulmonic flow velocities.

Range m/sec	peak +/- SD m/sec	Type	Transducer placement/ sample vol./angle corr.	Nr. of dogs	Sedation	Reference
0.76 - 1.22	0.98 +/- 0.1	CW	R./n.i./ n.i.	n=20		Yuill
	1.00 +/- 0.15	pulsed		n=28		Gaber
0.34 - 1.29	0.84 +/- 0.17	pulsed	R.para.short/PA/no	n=28	YES	Brown
0.88 - 1.61	1.20 +/- 0.20	pulsed	R para.short/PA/yes	n=50	no	Kirberger
0.43 - 1.38	0.95 +/- 0.19	pulsed	R para.short/RVOT/yes	n=50	no	Kirberger
0.60 - 1.91	1.25 +/- 0.26	CW	R.para.short/...../yes	n=50	no	Kirberger
< 1.3	1.07	pulsed				Bonagura
	0.99 +/- 0.22	n.i.	R or L cran/n.i./n.i.	n=20	no	Darke
0.88 - 1.34	1.06 +/- 0.14	pulsed	R para.short/PA/no	n=15	no	Bonagura et al
0.83 - 1.39	1.09 +/- 0.18	CW	R.para.short/PA/5-15°	n=24 (Boxers)	no	Le Bobinnec 1999 unpub

Mild stenosis:
peak gradient from 20 to 49 mmHg (2.25-3.5 m/sec)
Moderate stenosis:
peak gradient from 50 to 80 mmHg (3.5-4.5 m/sec)
Severe stenosis:
peak gradient above 80 mmHg (over 4.5 m/sec)

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