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Congenital Heart Disease (CHD) in dogs and cats

Dr. Djemai Samir

- Congenital heart diseases in cats and dogs (Congenital heart disease [CHD]) are:

❑ Either a malformation of one or more areas of the heart or large vessels (aorta or pulmonary artery).

❑ Or the persistence of a normal fetal structure after birth.

- Congenital heart diseases are relatively rare in general medicine for carnivores, but it is useful to know the most common diseases.

- Studies have shown that the prevalence of congenital heart diseases is about:

❑0.02% to 0.1% in cats.

❑0.5% and 1.0% in dogs.

- The table below lists the most common congenital diseases diagnosed in dogs and cats, in order of prevalence.

Most frequently reported congenital heart defects in dogs and cats, in order of prevalence.

Dogs	Cats
• Pulmonary stenosis • Patent ductus arteriosus • Aortic stenosis • Ventricular septal defect	• Ventricular septal defect • Aortic stenosis • Hypertrophic obstructive cardiomyopathy • Pulmonary stenosis

Ventricular Septal Defect (VSD)

Definition

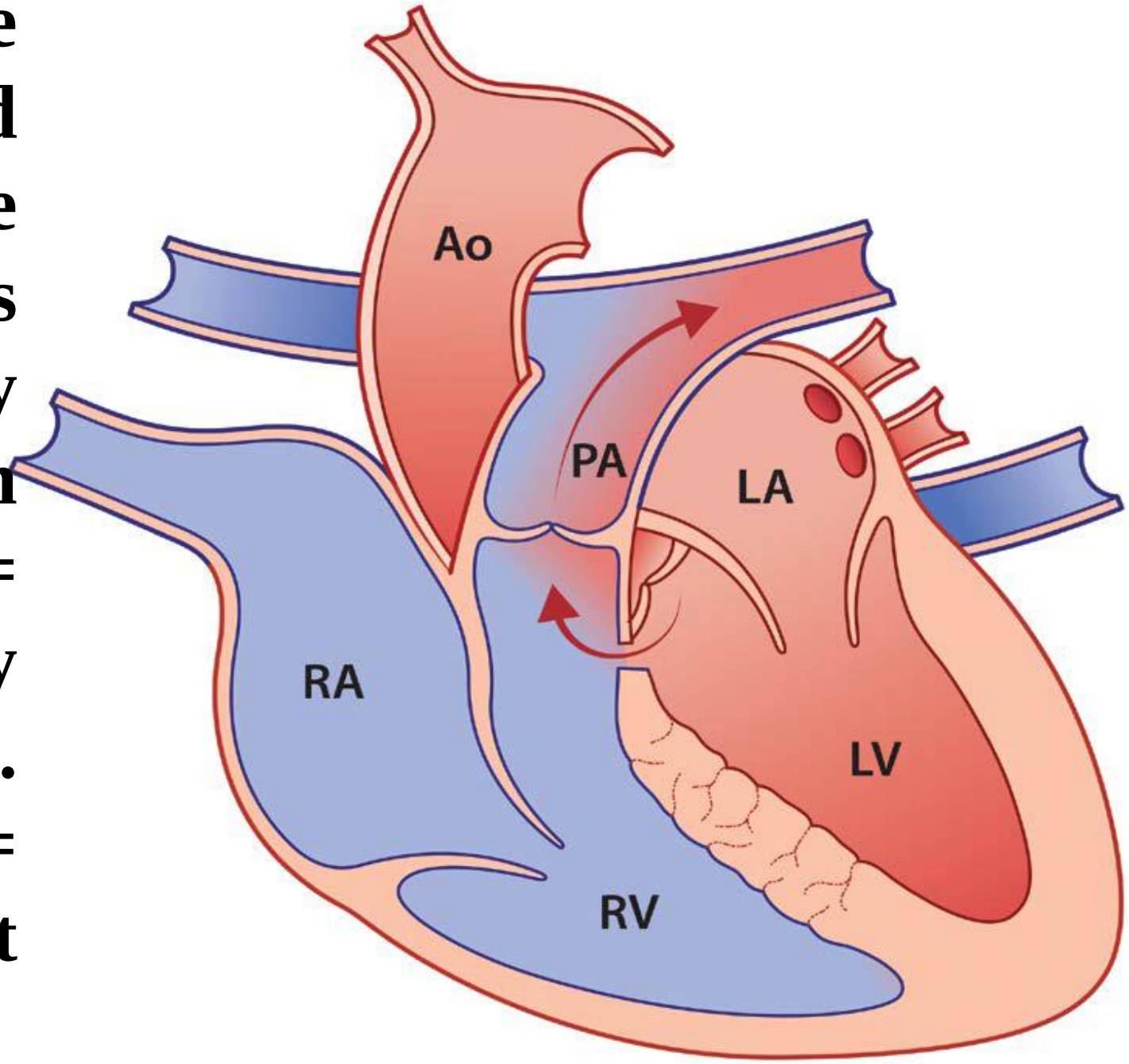
- The ventricular septal defect (VSD; Ventricular Septal Defect) is a congenital anomaly characterized by an opening (hole) in the wall of the interventricular septum allowing communication between the left and right ventricles.
- Generally asymptomatic = Asymptomatic CHD.
- The opening is usually located in the membranous part of the septum, just below the aortic valve.
- This is the most common congenital heart anomaly in cats.

Pathophysiology

- Due to higher pressures in the left ventricle, blood typically flows from the left side to the right side during systole = left-to-right shunt.
- This shunt (diversion) of blood is the cause of a heart murmur.

- Due to the location of the VSDs in the upper part of the outflow tract, the increased blood flow almost immediately enters the pulmonary artery and returns to the left heart.
- As a result, VSDs in dogs and cats generally lead to volume overload of the left ventricle and left atrium.

Picture of a VSD. Note how oxygenated blood from the left ventricle crosses the hole and flows into the pulmonary artery, mixing with deoxygenated blood. Ao = Aorta. Pa = Pulmonary artery. LA = left atrium. LV = Left ventricle. RA = Right atrium. RV = Right ventricle



- The outcome of a VSD depends on the size of the communication and the pressure gradient between the systemic circulation and pulmonary circulation.
- Very small VSDs (reduced blood flow) have minimal hemodynamic consequences.

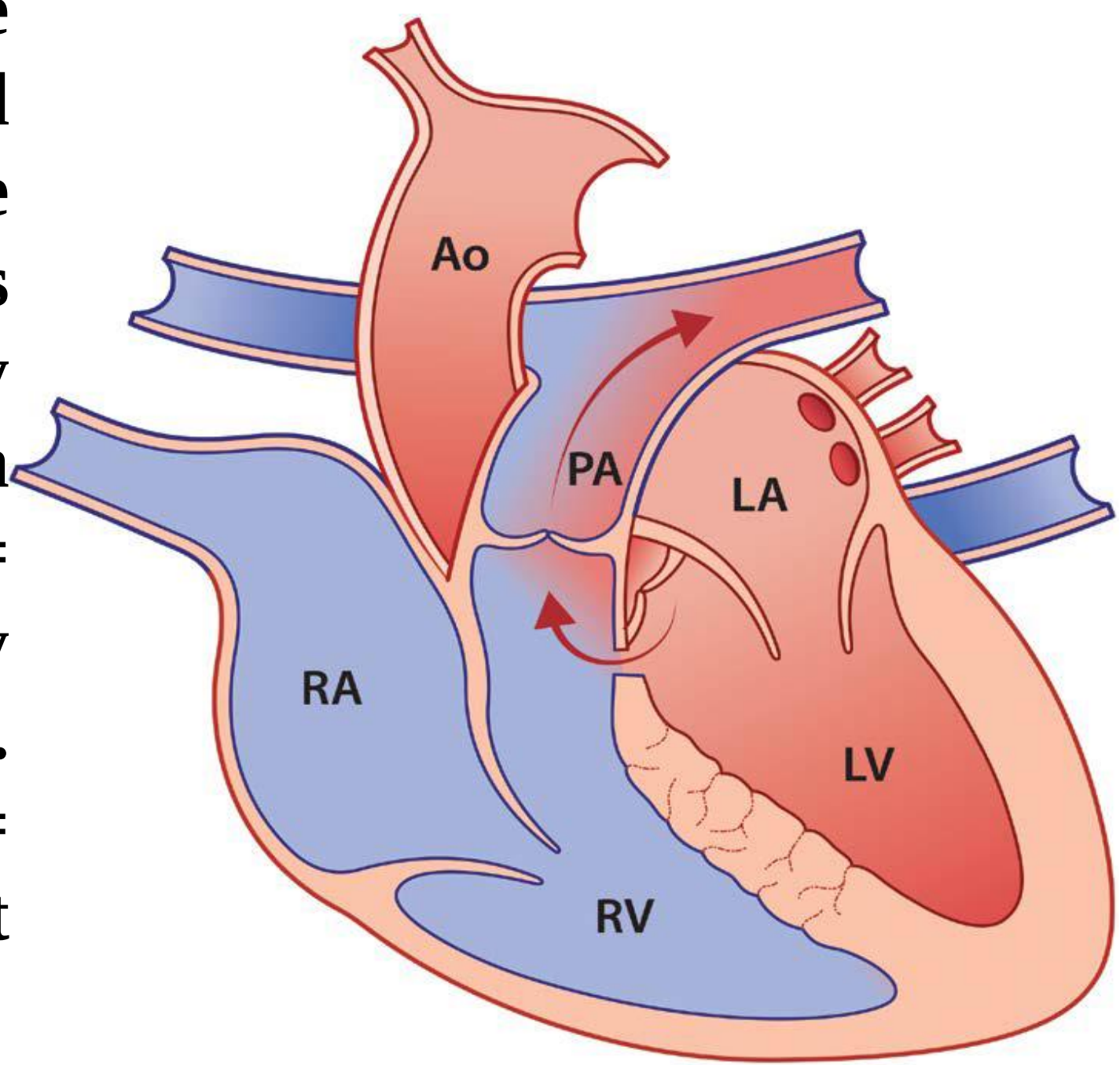
- Large VSDs:

❑Can lead to left-sided volume overload and subsequently left-sided heart failure (CHF).

❑Can also cause significant pulmonary overcirculation and lead to pulmonary hypertension (PH; Pulmonary hypertension).

❑In the presence of severe PH = pulmonary pressure (right heart) exceeds systemic pressure (left heart), reversing blood flow: deoxygenated blood enters the systemic circulation [right ventricular pressure exceeds left ventricular pressure, leading to a reversal of the shunt (right to left shunt) and cyanosis (Eisenmenger syndrome)].

Picture of a VSD. Note how oxygenated blood from the left ventricle crosses the hole and flows into the pulmonary artery, mixing with deoxygenated blood. Ao = Aorta. Pa = Pulmonary artery. LA = left atrium. LV = Left ventricle. RA = Right atrium. RV = Right ventricle

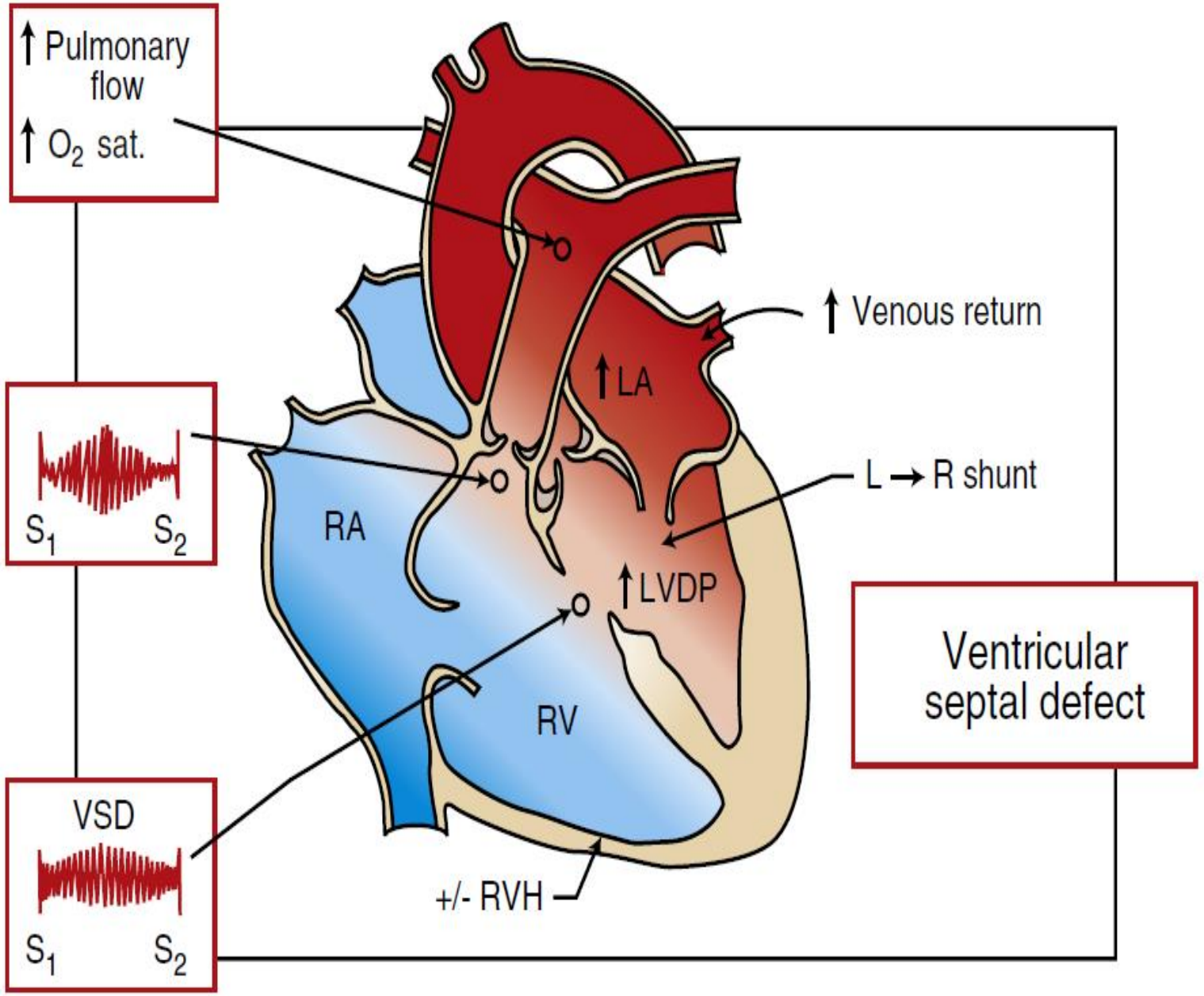


Clinical presentation

- The results of the physical examination depend on the size of the VSD (magnitude of the shunt) and the pressure gradient between the pulmonary circulation and the systemic circulation.
- Left-to-right shunts cause a right sternal systolic murmur; or left basal = depending on the location of the communication on the interventricular septum.

- Smaller VSDs often lead to a higher blood flow velocity (more resistance to blood flow) across the opening causing a louder murmur, but are often associated with fewer secondary changes.
- A larger hole offers less resistance, resulting in a softer murmur (quiet; or absence of audible murmur).
- Signs associated with CHF or hypoxemia may be present.

Pathophysiology and genesis of clinical findings in VSD. LA, Left atrium; LVDP, left ventricular developed pressure; RA, right atrium; RV, right ventricle; RVH, right ventricular hypertrophy.



Differential diagnosis

- Mitral insufficiency
- Tricuspid insufficiency
- Subvalvular aortic stenosis
- Pulmonic stenosis

Diagnosis

- The diagnosis is established by visualizing the interventricular communication (for example, 2D echocardiography) or the shunt with color Doppler echocardiography.
- At echocardiography, the size of the VSD can be compared to the diameter of the aortic ring:

❑Small VSDs represent less than 25% of the aortic diameter.

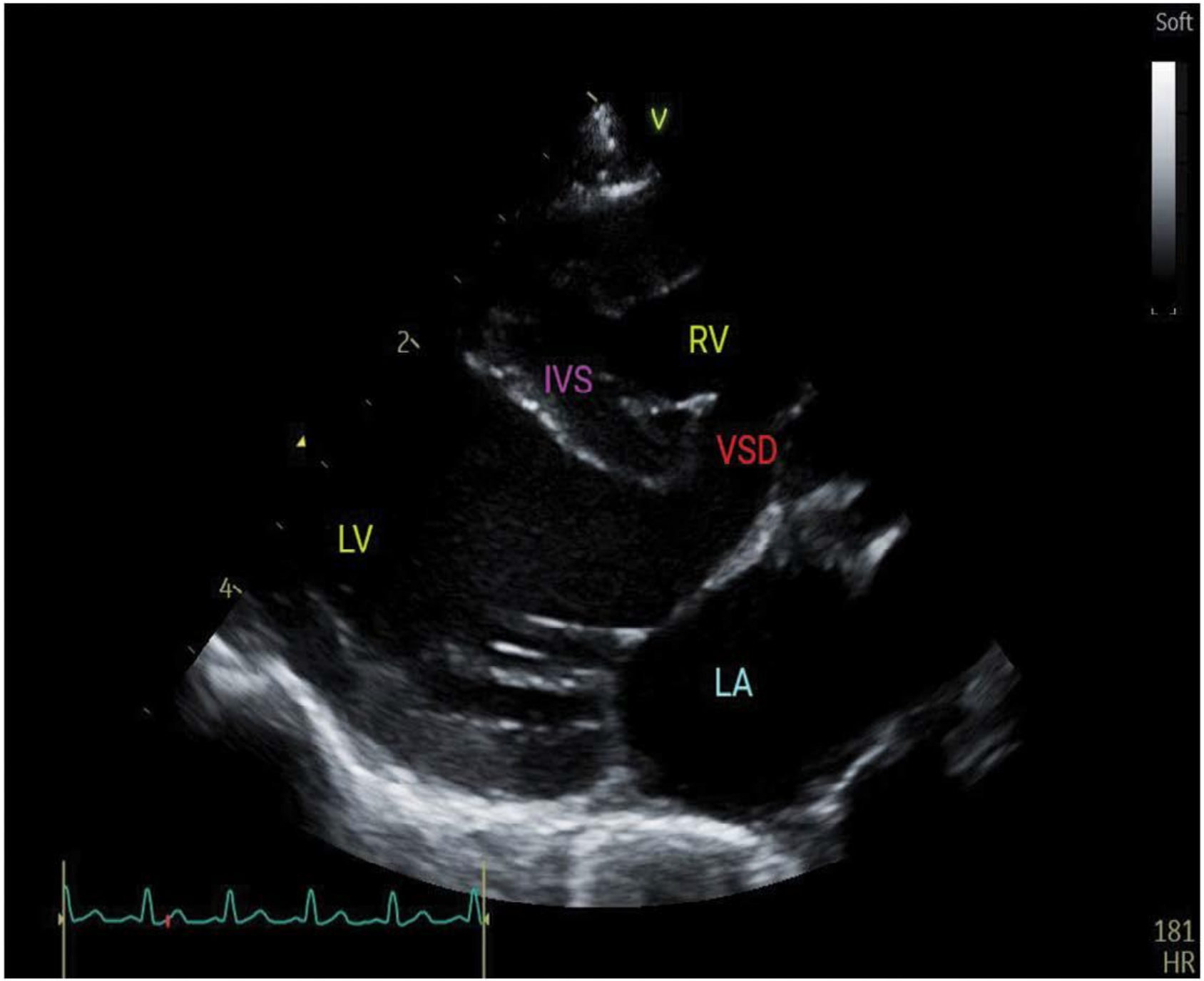
❑Moderate VSDs range from 25% to 75% of the aortic diameter.

❑Large holes (CIV) represent more than 75% of the aortic diameter.

- This comparison is necessary to determine the long-term hemodynamic consequences of a CIV.

- The direction and speed of blood flow through the communications are recorded by Doppler imaging.
- Clinical significance is better determined by measuring the size of the left heart chambers = left atrium and left ventricle.

VSD on echo. IVS = Intraventricular septum, RV = right ventricle. RV = Right ventricle. LV = Left ventricle. IVS = Intraventricular septum. LA = Left atrium.



Management

- CIVs are managed based on the severity of secondary changes and clinical signs.
- Small CIVs may not require any intervention and have a good prognosis = Treatment is not recommended in this case.
- CIVs with hemodynamically significant left-to-right shunt are managed medically when signs of left heart failure appear.

- Cyanosis and polycythemia resulting from VSD with right-to-left shunt can improve with sildenafil, which targets HPT.

- Closure by surgery or catheter:

❑Open-heart techniques for closure or surgical repair of VSD are curative; however, these techniques are available in very limited locations and are often very expensive.

❑Palliative reduction of the shunt can be achieved by banding the pulmonary artery = leading to an increase in right ventricular systolic pressure = When right ventricular pressure rises, the volume of the shunt decreases and pulmonary circulation is spared the deleterious effects of chronic volume overload.

❑Catheter closure of the VSD with an occlusion device is described = but technically difficult.

Prognosis

- Prognosis varies depending on the size of the VSD.

Patent ductus arteriosus (PDA)

Definition/overview

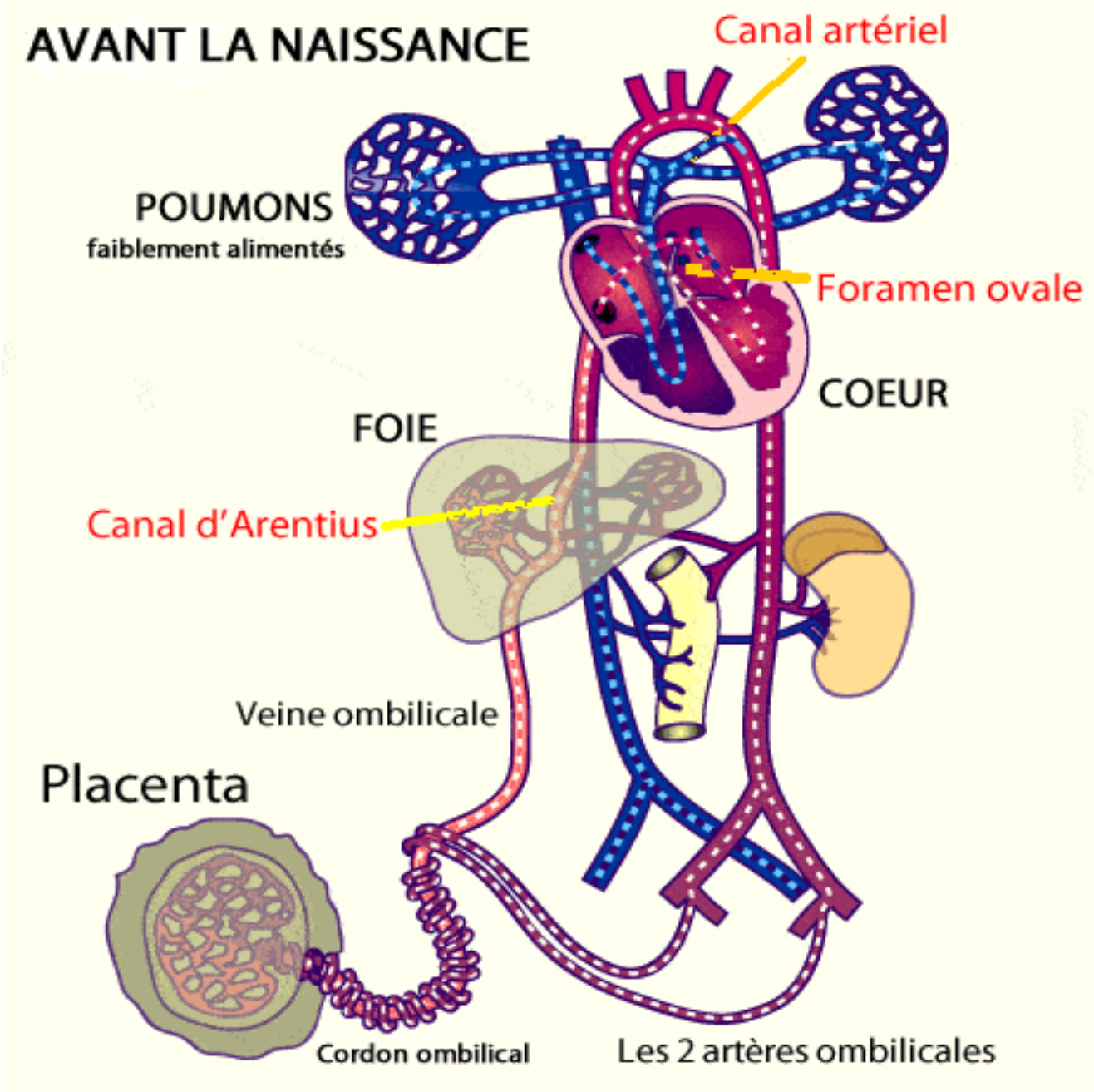
- Persistence of the ductus arteriosus (PDA; Patent ductus arteriosus [PDA]) is a congenital heart defect in which the normal ductus arteriosus does not close after birth:

❑In utero (in the fetus), the ductus arteriosus connects the aorta and the pulmonary artery; it allows blood to bypass the lungs.

❑Normally, the ductus closes 7 to 10 days after birth.

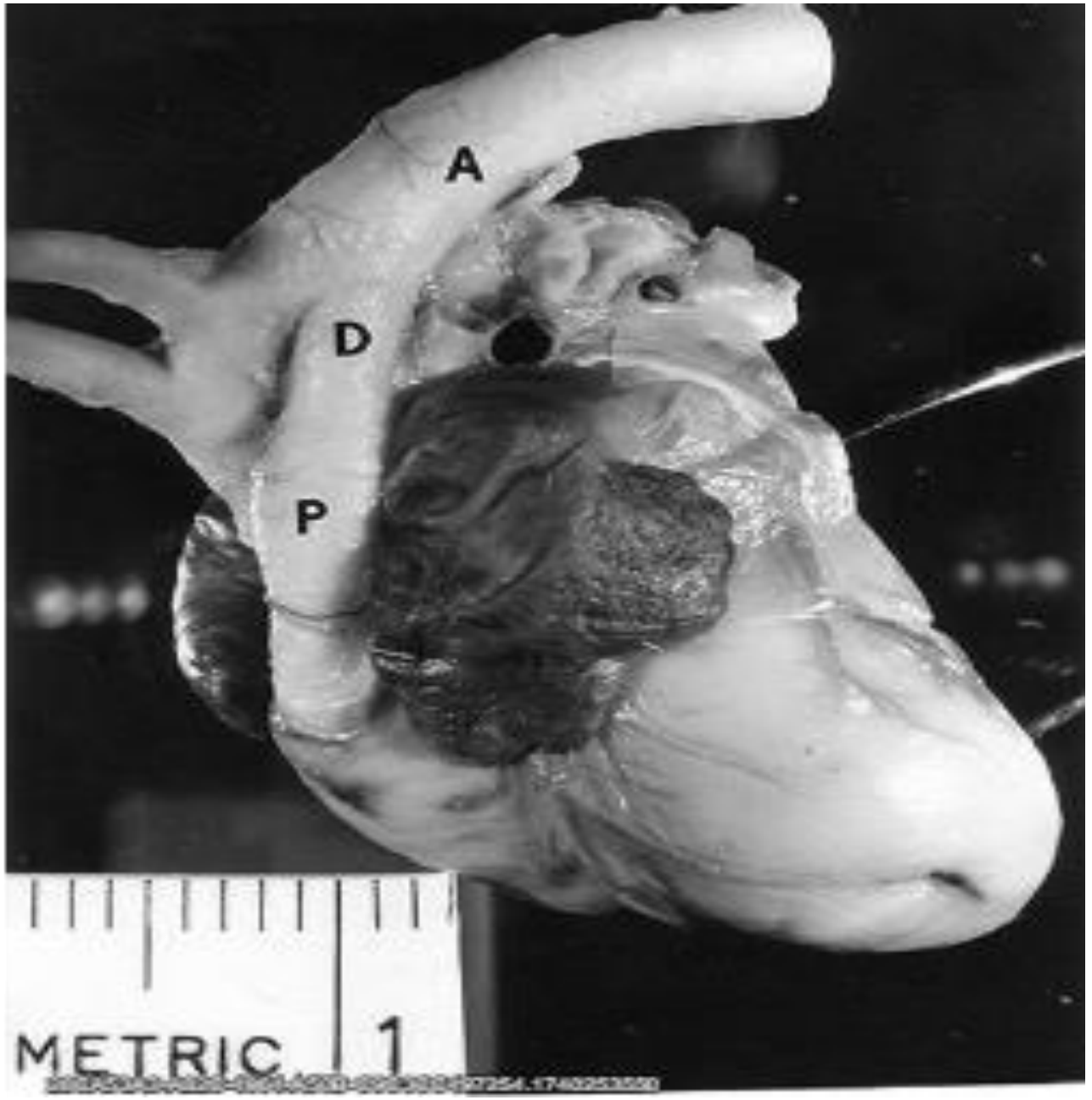
❑In some rare cases, the ductus arteriosus remains open (patent) beyond this period, allowing blood to flow depending on the pressure gradient.

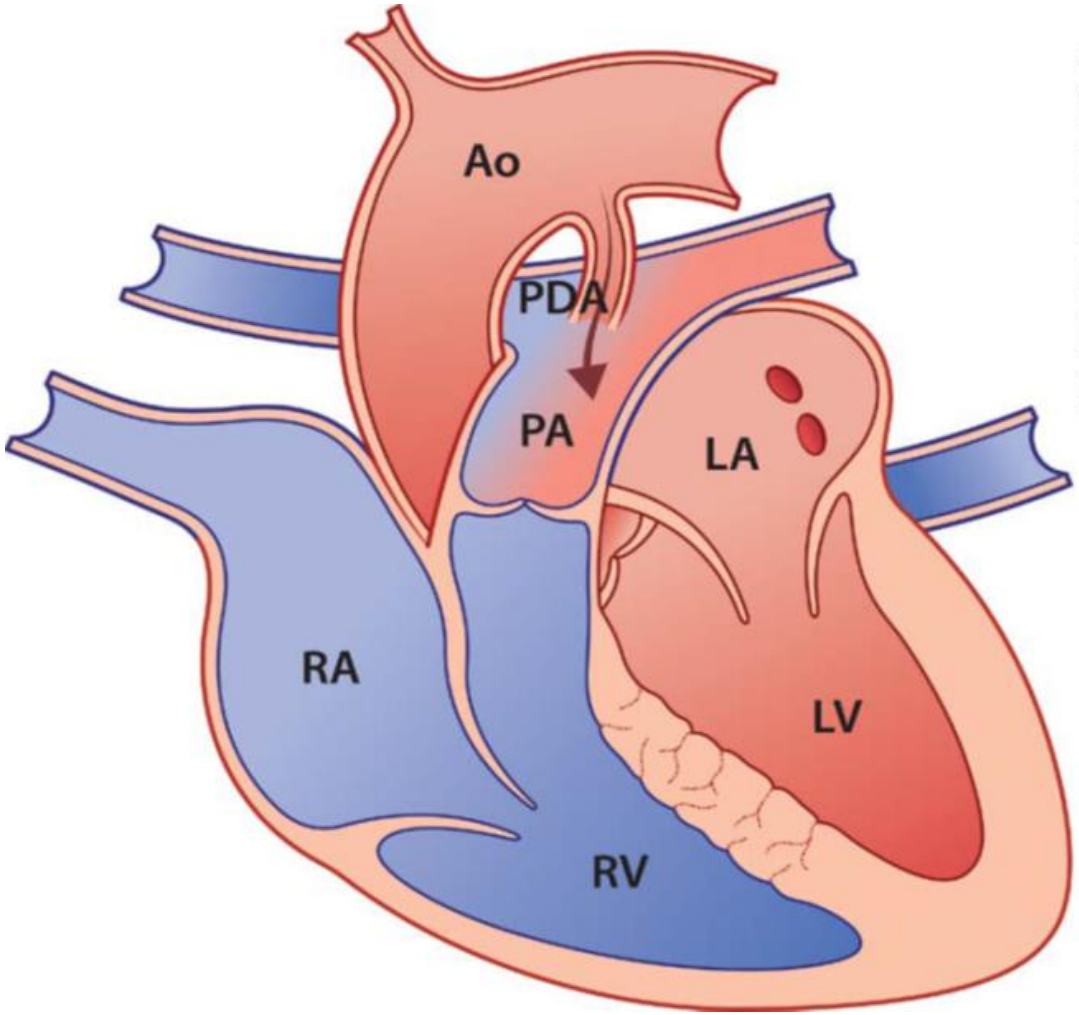
The ductus arteriosus is a blood vessel, functional only in the fetus, that connects the pulmonary trunk to the aorta and allows blood to bypass pulmonary circulation. Indeed, fetal blood oxygenation occurs via the placenta and not through the lungs.



Photograph of the heart and large vessels in a normal newborn dog.

The ductus arteriosus (D) is partially narrowed; it extends between the pulmonary artery (P) and the aorta (A).





Ao = Aorta
PA = Pulmonary artery
LA = Left atrium
LV = Left ventricle
RA = Right atrium
RV = Right ventricle
PDA = Patent ductus arteriosus

Persistence of the ductus arteriosus (PDA)
Patent ductus arteriosus (PDA)

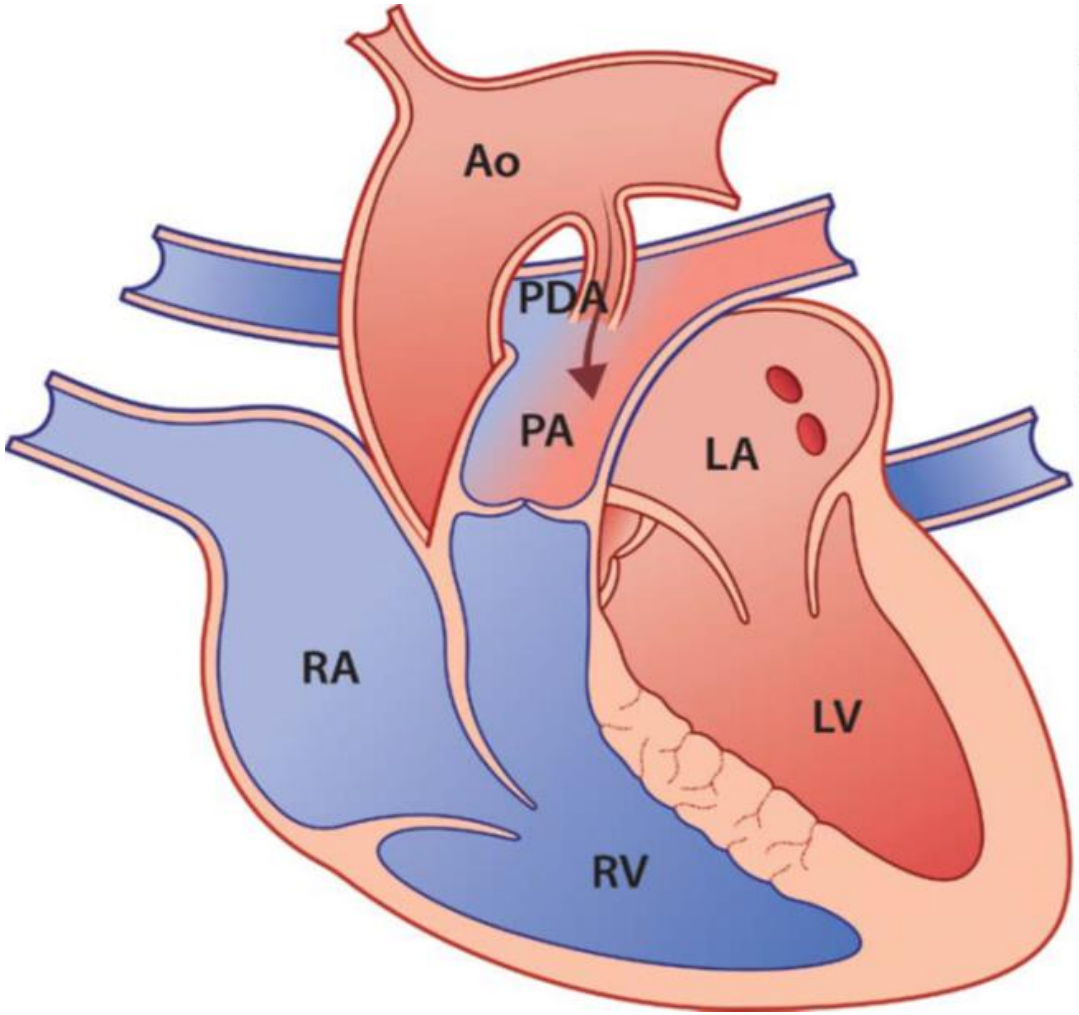
- There seems to be a racial predisposition, particularly in German Shepherds, Shetland Sheepdogs, English Springer Spaniels, Bichon Frise, Poodles, and Yorkshire Terriers.
- A larger number of affected dogs are females.
- Cats can also be diagnosed with a PCA = but it is less common.

Bichon frisé

Poodle

Pathophysiology

- Intracardiac pressure in the aorta is higher than in the pulmonary artery = In most cases, a PCA leads to a blood shunt from the aorta to the pulmonary artery (left-to-right shunt) and volume overload on the left side.
- Blood that goes directly to the pulmonary circulation returns to the left side of the heart.
- As a result, the left ventricle and left atrium dilate.
- Significant shunts increase the filling pressure in the left heart and pulmonary veins, leading to left-sided congestive heart failure (CHF).
- The severity of the shunt is primarily determined by the size of the permeable vessel (canal).



Ao = Aorta
PA = Pulmonary artery
LA = Left atrium
LV = Left ventricle
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RV = Right ventricle
PDA = Patent ductus arteriosus

Persistence of the arterial duct (PCA)
Patent ductus arteriosus (PDA)

- In rare cases with significant shunts and severe pulmonary arterial hypertension (HTP), shunt reversal is observed:

❑Blood flows from the pulmonary artery to the aorta – a right-to-left shunt – causing differential cyanosis, hypoxemia, and ultimately polycythemia (Eisenmenger syndrome).

❑Shunt reversal typically occurs in puppies aged a few days to a few weeks.

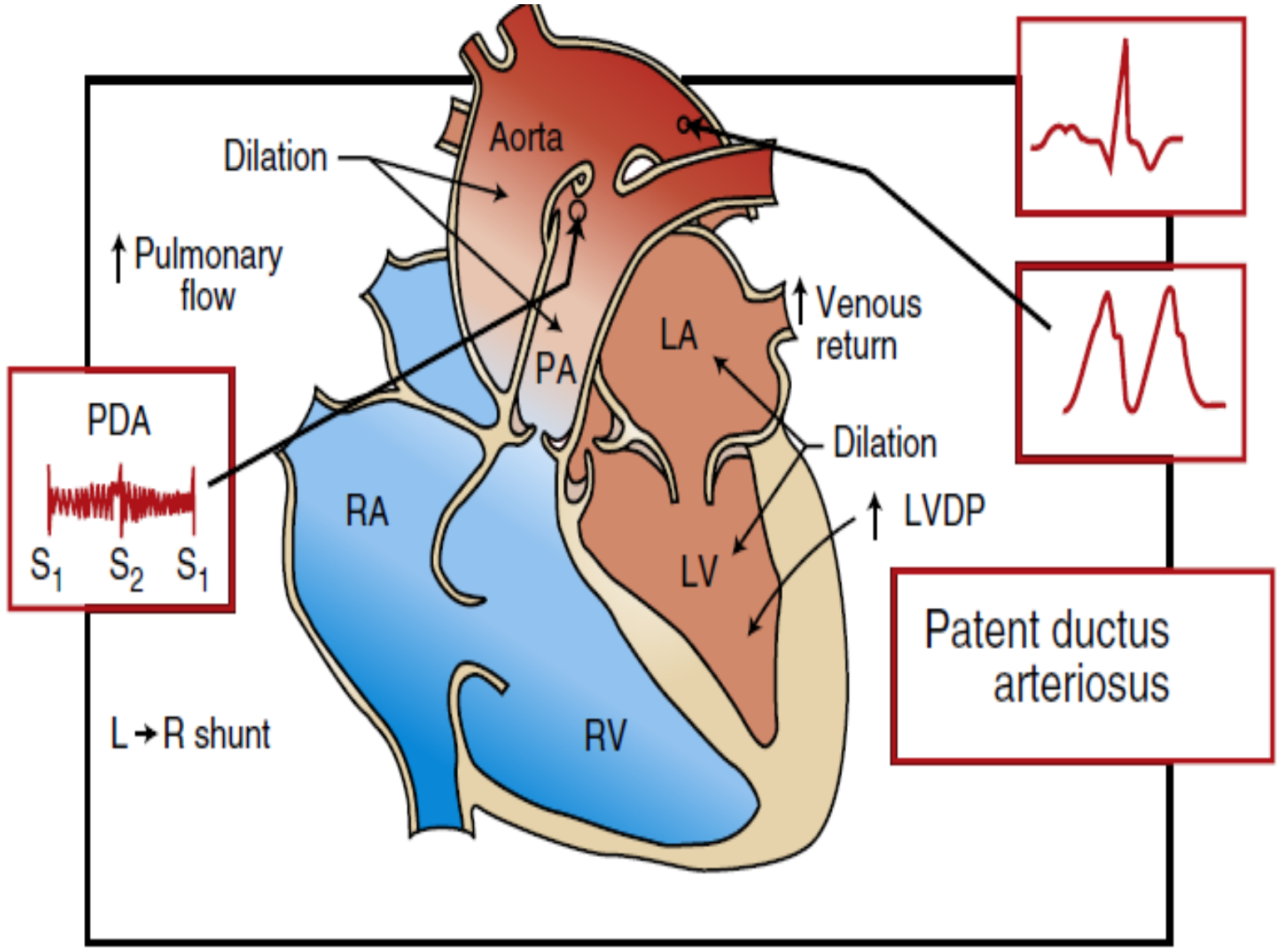
❑This is not the usual progression of a PCA in adult dogs.

Clinical presentation

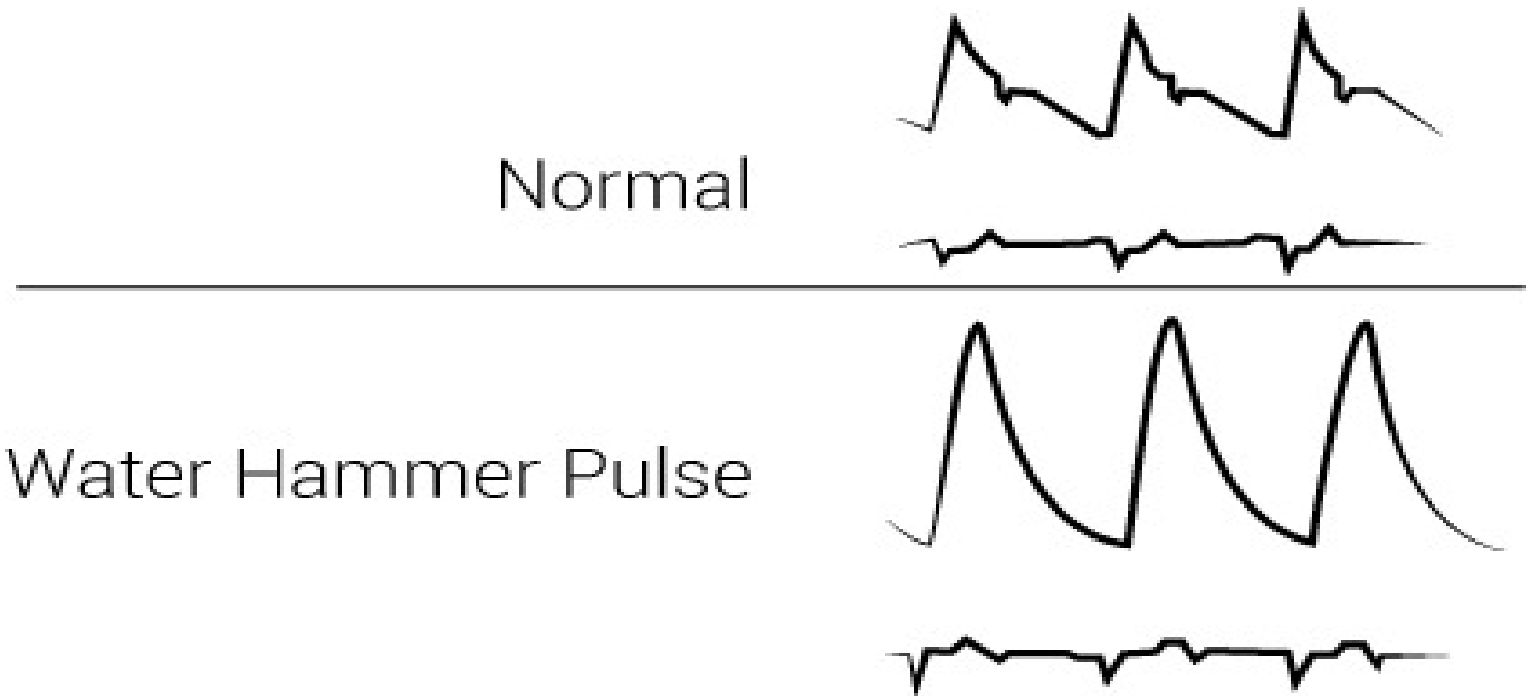
- Clinical signs vary depending on the size of the shunt, its direction, and the presence of heart failure.
- Signs may include exercise intolerance, syncope, cough or dyspnea, and in the case of a reversed shunt, weakness of the hind limbs or seizures.
- Upon physical examination of animals with a PCA with left-to-right shunt, a high-grade, continuous ('machinery') murmur is heard cranially in the left axillary region.
- The murmur increases during systole and decreases during diastole.

- A strong cardiac murmur (grade V/VI or VI/VI) is often accompanied by a thrill that can be palpable at the cranial thorax.

Pathophysiology and genesis of clinical findings in patent ductus arteriosus (PDA). LA, Left atrium; LV, left ventricle; LVDP, left ventricular developed pressure; PA, pulmonary artery, RA, right atrium; RV, right ventricle.



- Bounding femoral pulses (hyperdynamic = water-hammer pulse) can be palpated due to the large difference between systolic and diastolic pressures.



- In animals with a reverse PCA, there is no audible murmur, but a careful inspection of the mucous membranes should reveal differential cyanosis, meaning cyanosis limited to the caudal mucous membranes.

Differential diagnosis

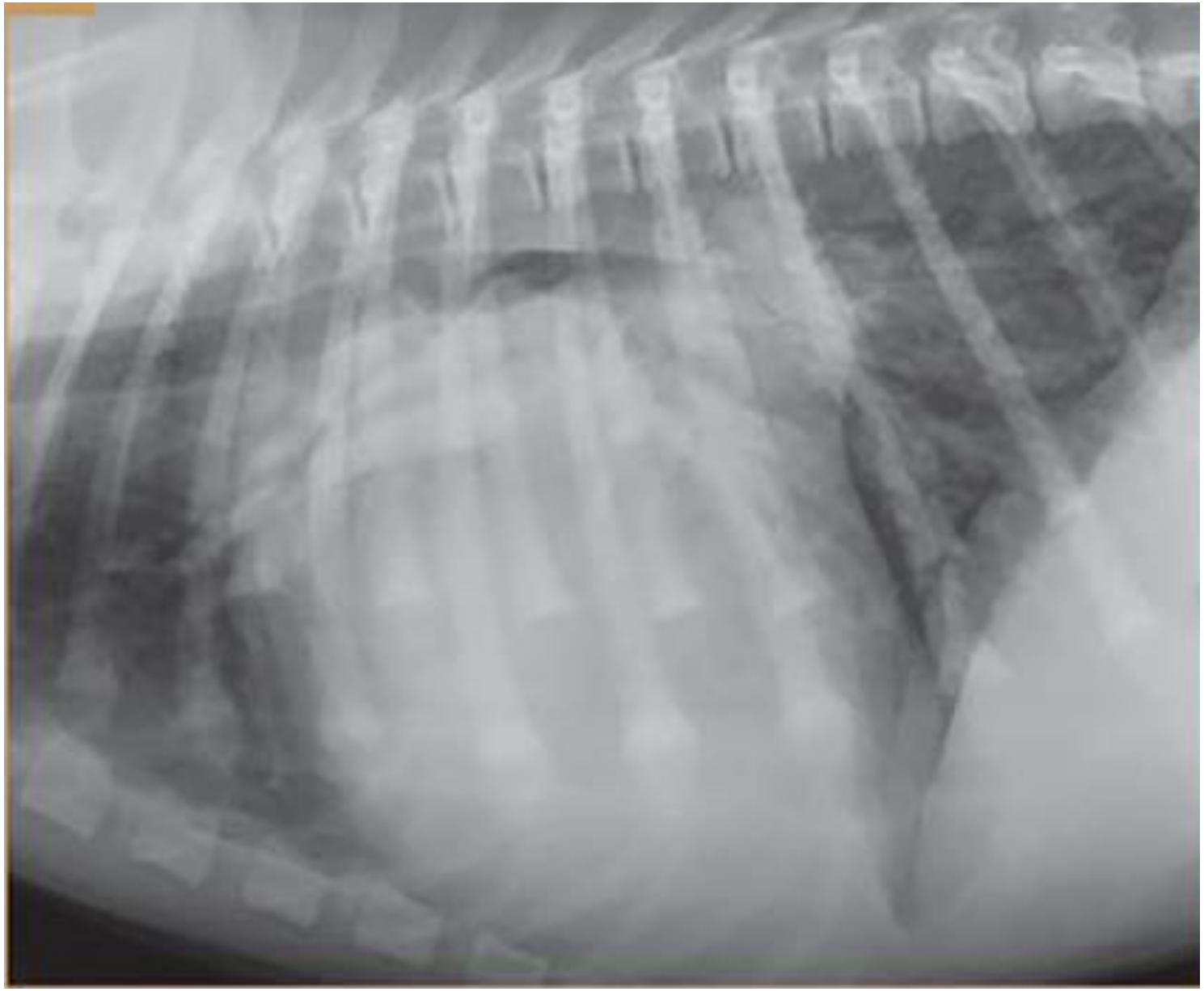
- Aortic stenosis.
- Ventricular septal defect.

Diagnosis

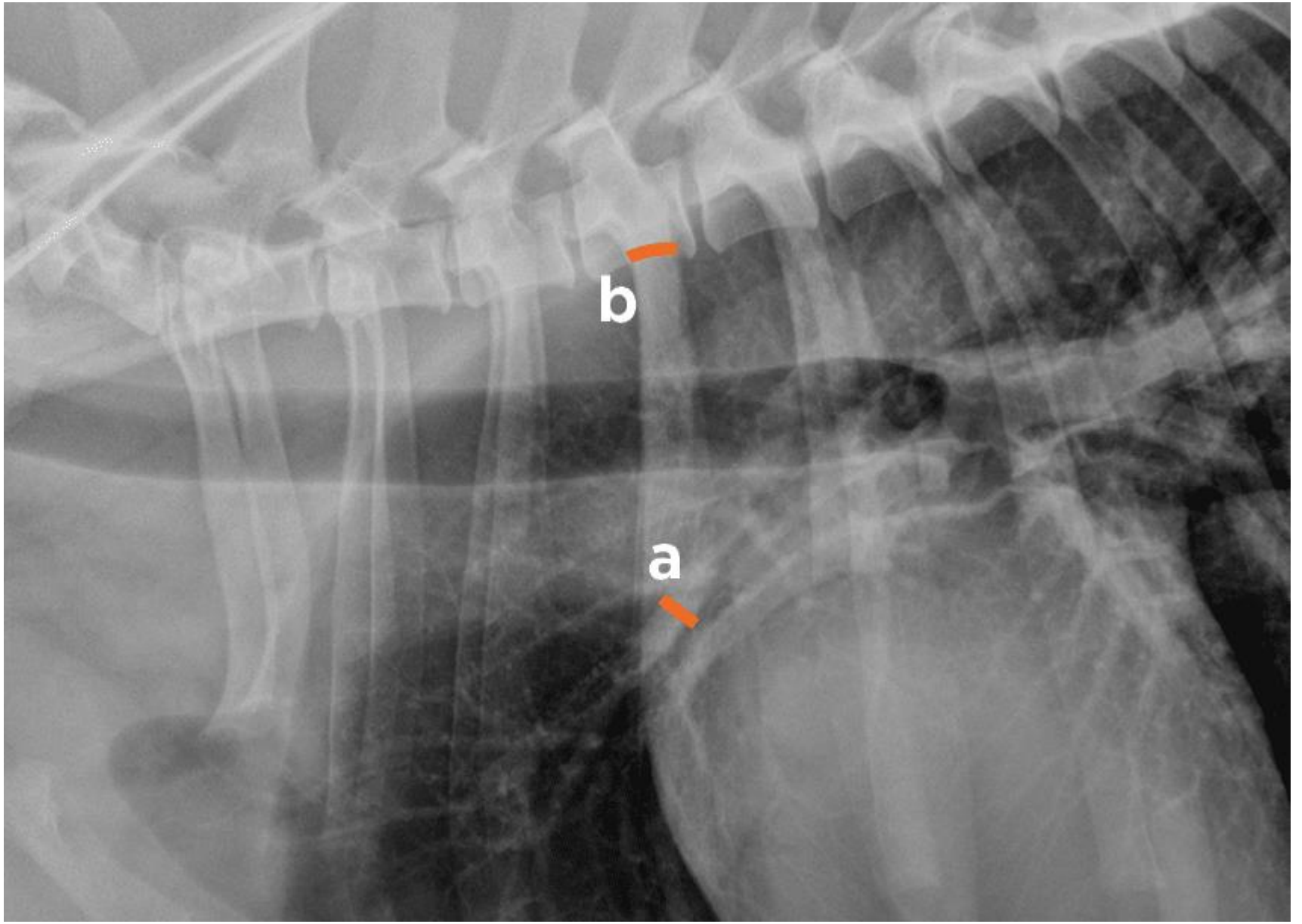
Chest X-rays may show:

- Left cardiomegaly (left atrial and ventricular dilation).
- Dilated pulmonary trunk related to pulmonary overcirculation (pulmonary overcirculation).
- Dilated descending aorta.
- Aneurysm (dilation) of the aorta at the insertion of the ductus.
- Left heart failure sign = Pulmonary edema sign (±).
- Right cardiomegaly in the case of an inverted PCA.

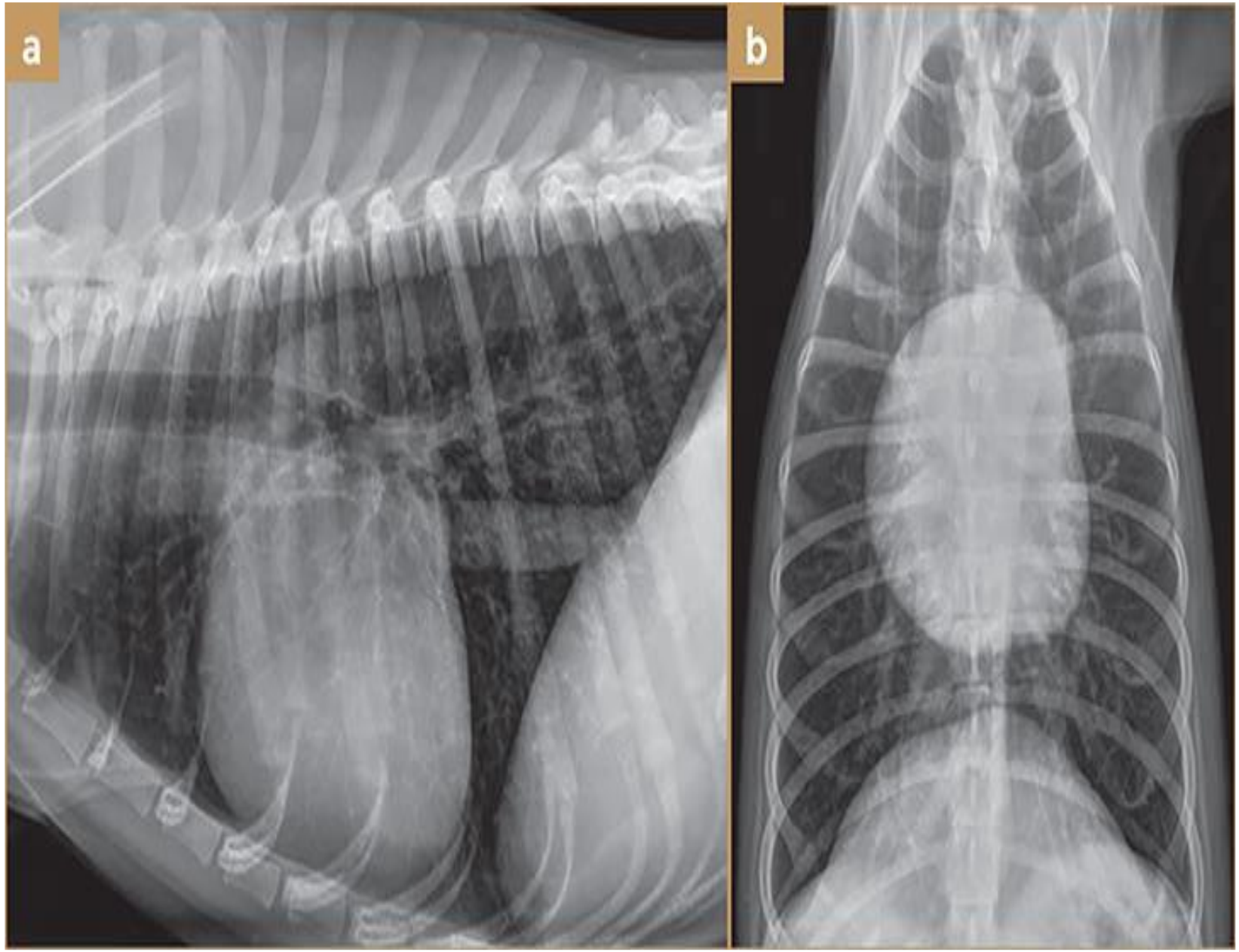
Six-month-old female German shepherd. Left lateral view; severe cardiomegaly with severe dilation of the left ventricle and atrium and the cranial lobar pulmonary veins and arteries, with congestion of the pulmonary hilum and diffuse interstitial infiltration.



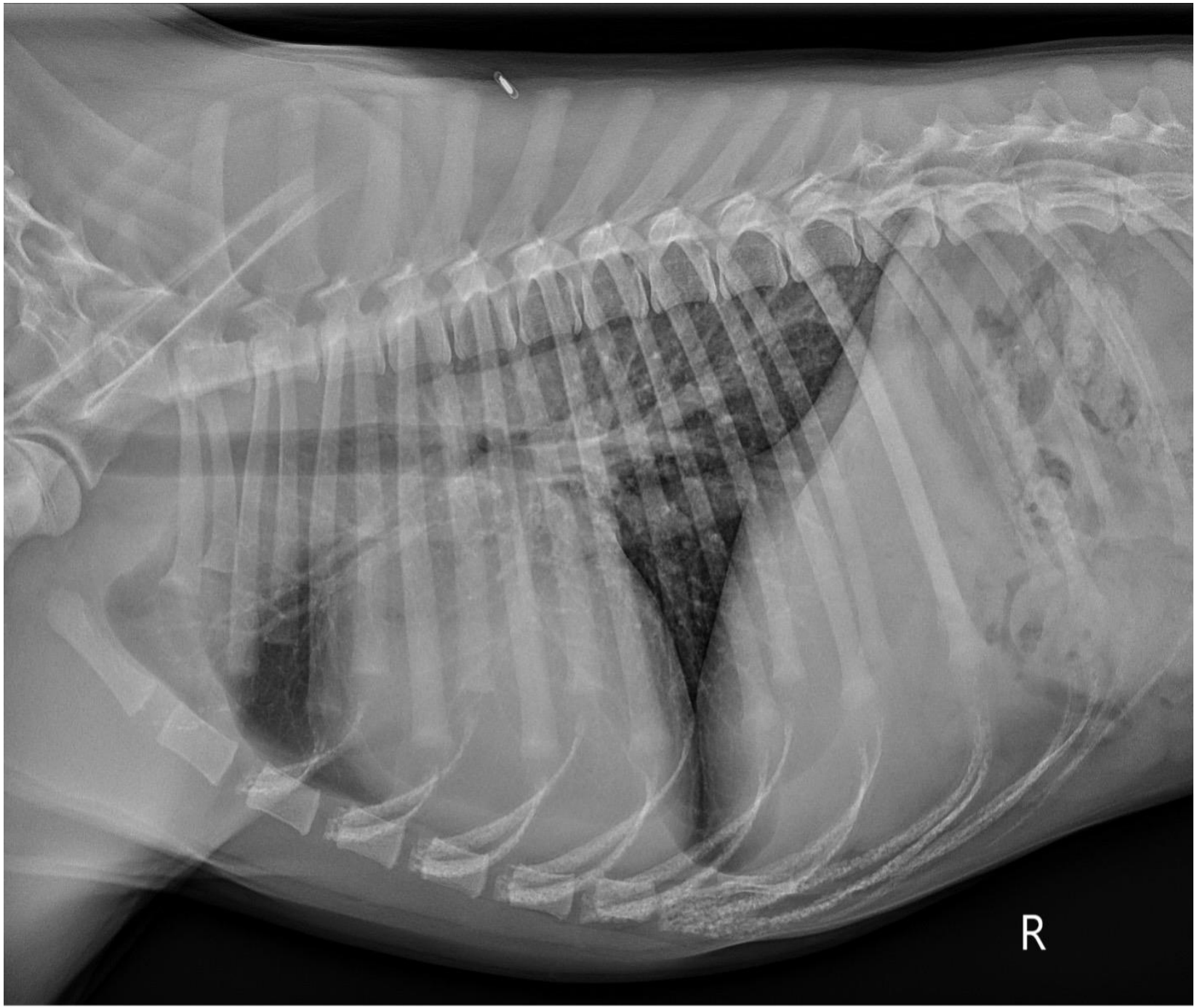
Left lateral projection of a dog without cardiovascular disease and (a) a normal right cranial lobar pulmonary artery width compared to (b) fourth rib width.



One-year-old female Collie showing an inverted PCA and an increase in the size of the right ventricle. a) Increased sternal contact on the lateral view. b) Ventrodorsal view; cardiac silhouette in the shape of an inverted D.



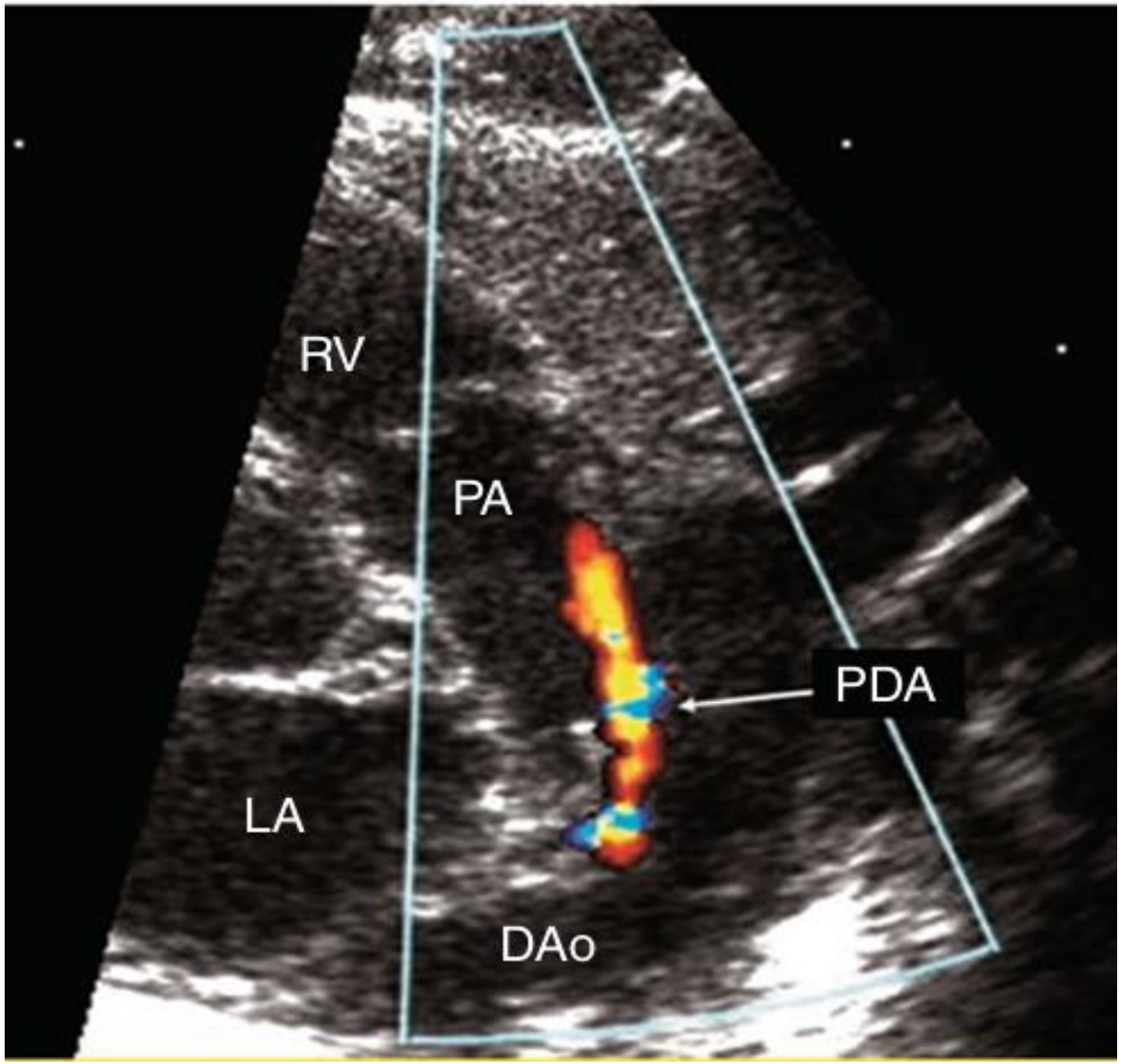
Chest radiographs: marked cardiomegaly with pulmonary vascular overcirculation. A slight diffuse bronchial pattern is noted.



Echocardiography (2D, M-mode, and Doppler) is necessary to visualize the channel and confirm the presence of the shunt.

- Left atrial and ventricular dilation
- Assess the severity of the enlargement of the left cardiac chambers.
- Dilation of the pulmonary artery.
- Identify concomitant coronary heart diseases.
- Confirm the presence of mild to severe pulmonary hypertension (HTP).
- Mitral regurgitation due to left heart dilation and stretching of the mitral annulus (color Doppler).
- Tricuspid regurgitation due to right heart dilation associated

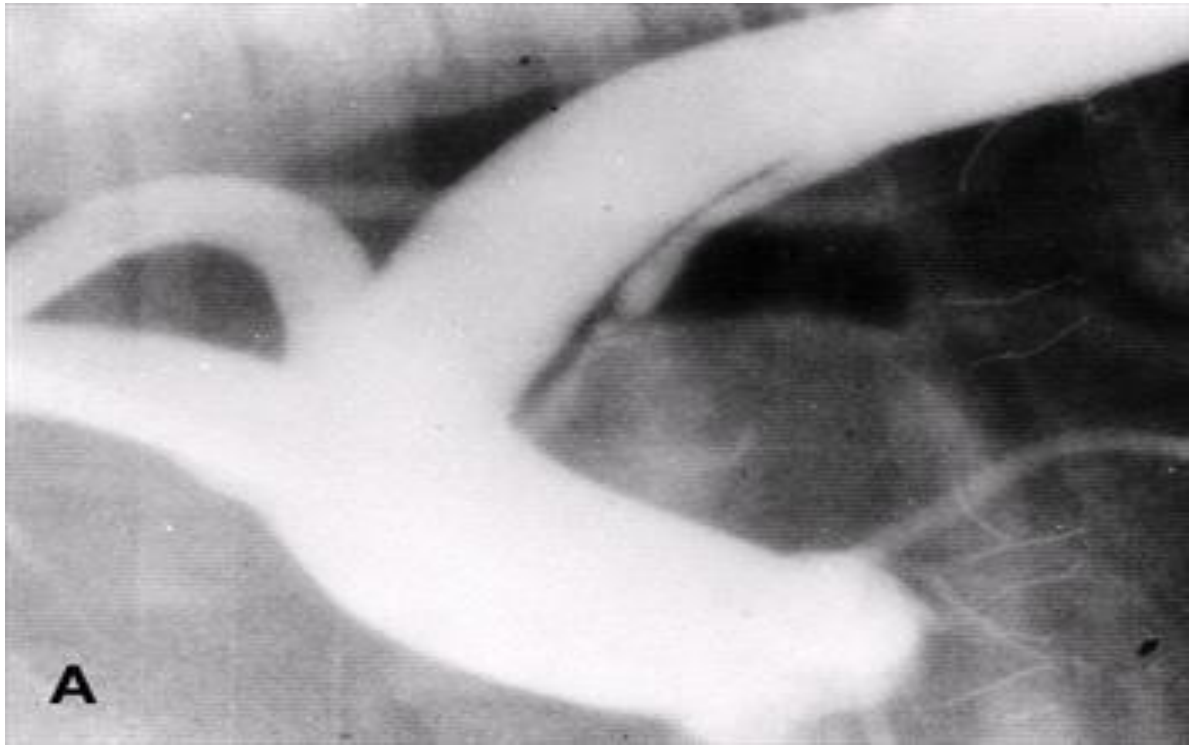
Color Doppler image view of a patent ductus arteriosus (PDA). PA, pulmonary artery; PDA, patent ductus arteriosus; Ao, aorta; LA, left atrium, RV, RV, right ventricle.



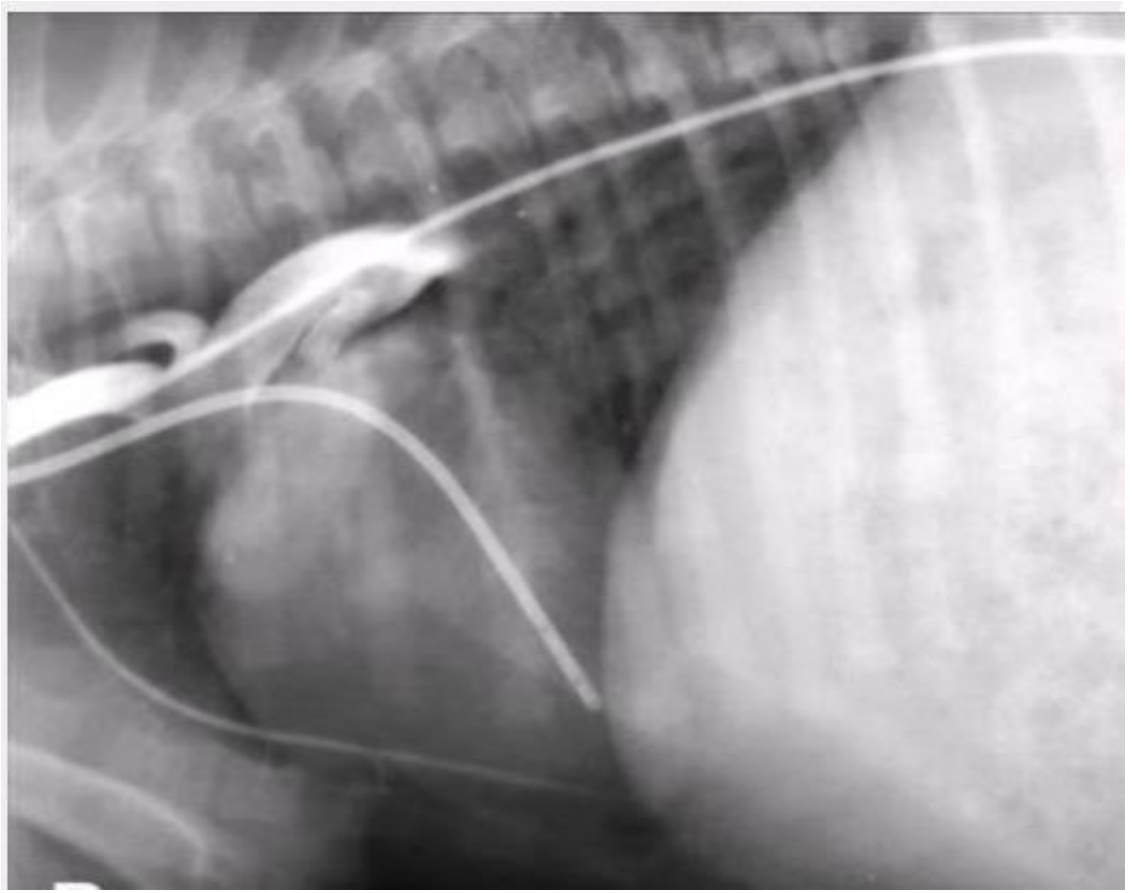
Electrocardiography may show:

- ❑May be normal.
- ❑Frequently an increased amplitude of the R wave (tall R wave) in lead II = Indicating left ventricular dilation.
- ❑In reversed PCA: Deep Q waves in leads I, II, III, and avF + right axis deviation (to the right).
- ❑Wide P waves (duration).
- ❑Various arrhythmias, particularly atrial fibrillation.

- ***Cardiac catheterization and angiography = Visualize the PCA, assess pressures, and determine the direction of flow through the PCA.***



Aortography showing a left-to-right shunt (L-R).



Presence of a tubular pulmonary arterial canal of medium size with a G-D shunt

Treatment

- If heart failure and arrhythmias are present, they should be treated before the canal is occluded.
- Management of heart failure:

❑ Diuretics (furosemide, spironolactone).

❑ Pimobendan.

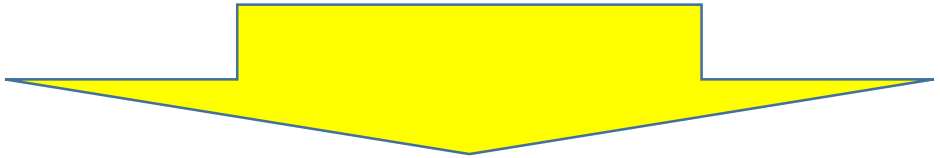
❑ ACE inhibitors.

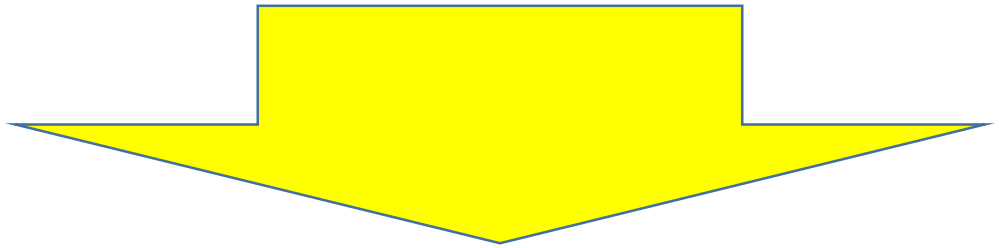
- Phlebotomies (to maintain hematocrit at 55–65 %) are indicated when polycythemia is responsible for clinical signs (usually for a hematocrit [packed cell volume (PCV)] >70 %).

- PCA are best treated by early closure (or occlusion) = particularly for the left-to-right shunt:

❑ Either by surgical ligation = surgical occlusion.

❑ Either by cardiac catheterization procedures = Occlusion by embolization.





Surgical ligation:

- Surgical ligation is the treatment of choice for PDA with left-to-right shunt.
- Small patients who are not sensitive to catheterization are better treated surgically.
- Surgery has a success rate of up to 95%

Occlusion by embolization = Cardiac catheterization procedures (Minimally invasive transcatheter procedures):

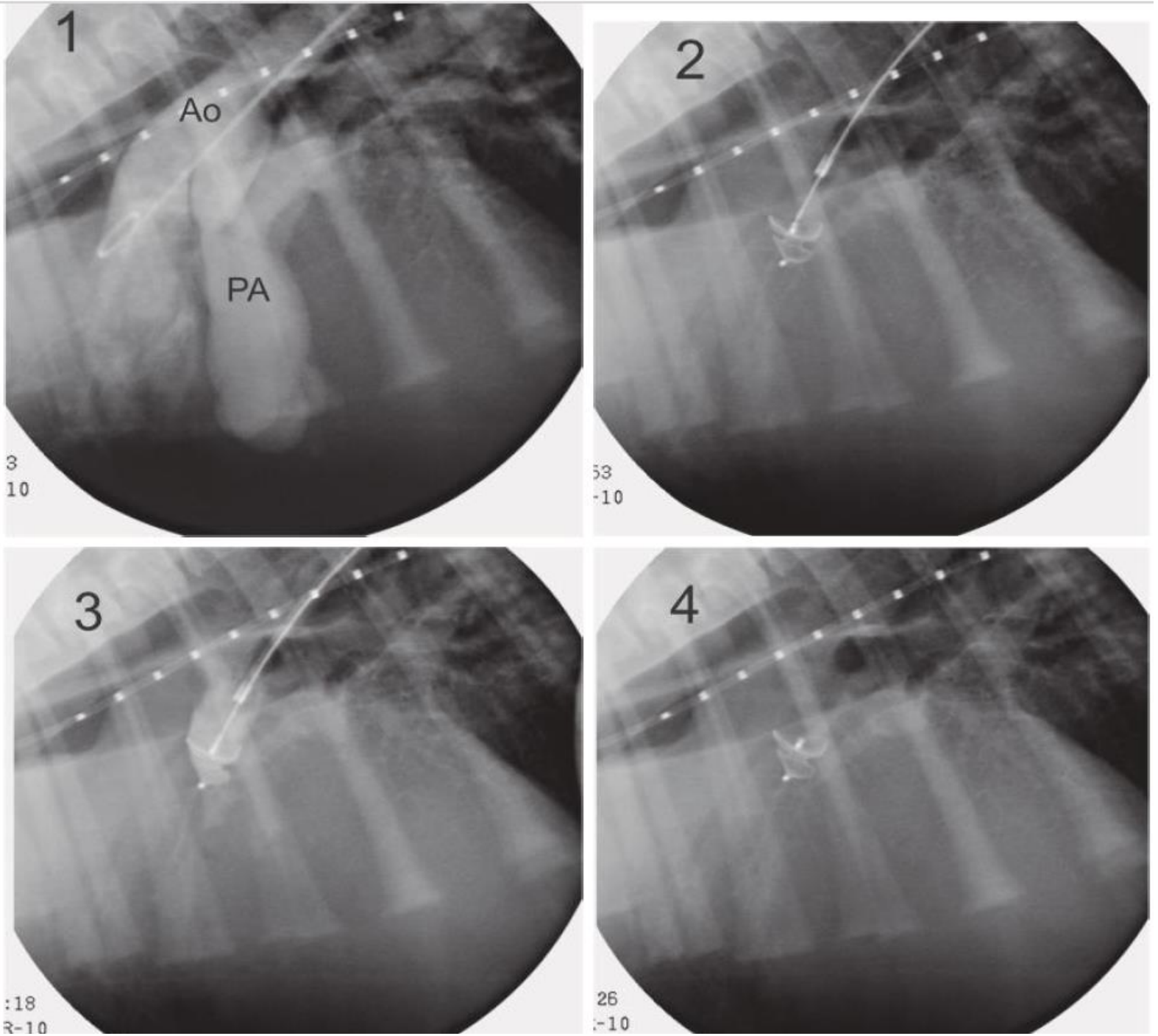
- The shunt through the ductus arteriosus can be stopped by a minimally invasive catheter occlusion device.
- Embolization using an arterial embolization implant to occlude the PDA = . This can be done using:
  - Vascular coils = Spirals (Coil) or coil.

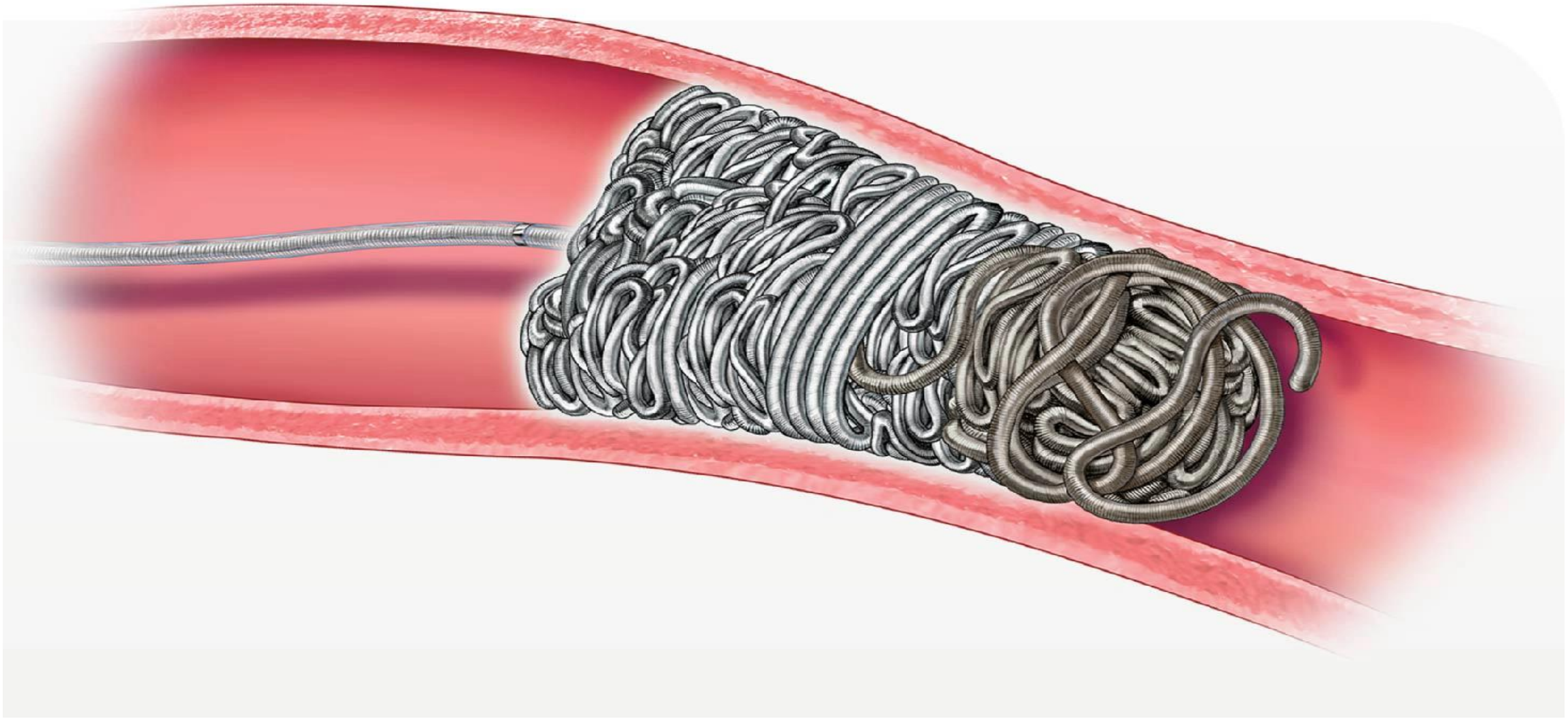
or

  - Vascular plugs : Amplatz (Amplatz canine ductal occluder, ACDO)
- This embolization can be performed via the trans-arterial route (the catheter is passed through the femoral artery) or transvenous (transvenous approach accessing the arterial duct on the side of the pulmonary artery).
- The trans-arterial route is the widely preferred route.
- This procedure uses the introduction of a coil or duct occlusion device into the lumen of the arterial duct, which leads to embolization of the arterial duct.

Arterial embolization coils (on the left) and Amplatz (on the right; Amplatz canine ductal occluder = it is placed via the femoral artery in the PDA, to close the duct).

Serial fluoroscopic images and angiocardiograms obtained during a catheter PCA occlusion procedure. 1) The angiogram reveals a funnel-shaped PCA and a left-to-right shunt from the aorta (Ao) to the pulmonary artery (PA). 2) The embolization device (attached to a catheter) is positioned on the PCA. 3) The angiogram reveals the occlusion of the PCA by the device. 4) The device is disconnected from the catheter and remains properly positioned on the PCA.





Coils (Coil, Vascular coils)

NB:

In the case of reversed PCA:

- The closure of reversed PCAs is contraindicated = reversed PCAs often cannot be safely closed.
- Sildenafil to reduce pulmonary pressures.

Prognosis

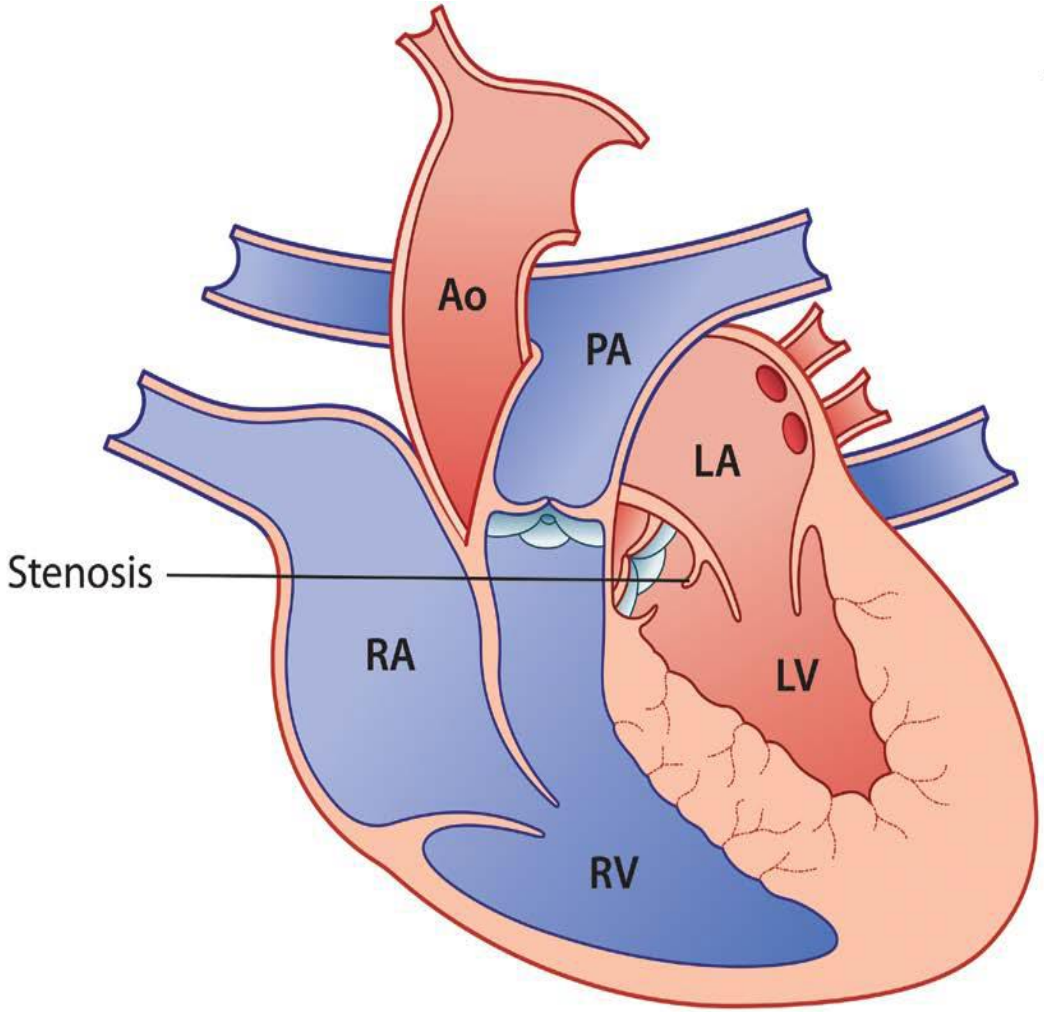
- 64% die before the age of one without intervention.
- Normal or nearly normal life expectancy if the patent ductus arteriosus is the only congenital anomaly and is closed before significant dilation and dysfunction of the left chambers develop.
- Surgery in older dogs carries a higher risk as the patent ductus arteriosus and the dilated pulmonary artery become fragile.

Aortic Stenosis (AS)

Definition

- Aortic stenosis results from a narrowing of the outflow tract of the left ventricle (left ventricular outflow tract [LVOT]) in the area of the aortic valve and can occur at the subvalvular, valvular, or supravalvular level.
- In dogs, the most common manifestation is Subvalvular Aortic Stenosis (Subvalvular aortic stenosis [SAS]).
- A genetic mutation responsible for SAS has been identified in Newfoundlands.

Aortic stenosis



Ao = Aorta
PA = Pulmonary artery
LA = Left atrium
LV = Left ventricle
RA = Right atrium
RV = Right ventricle
PDA = Patent ductus arteriosus

Newfoundland

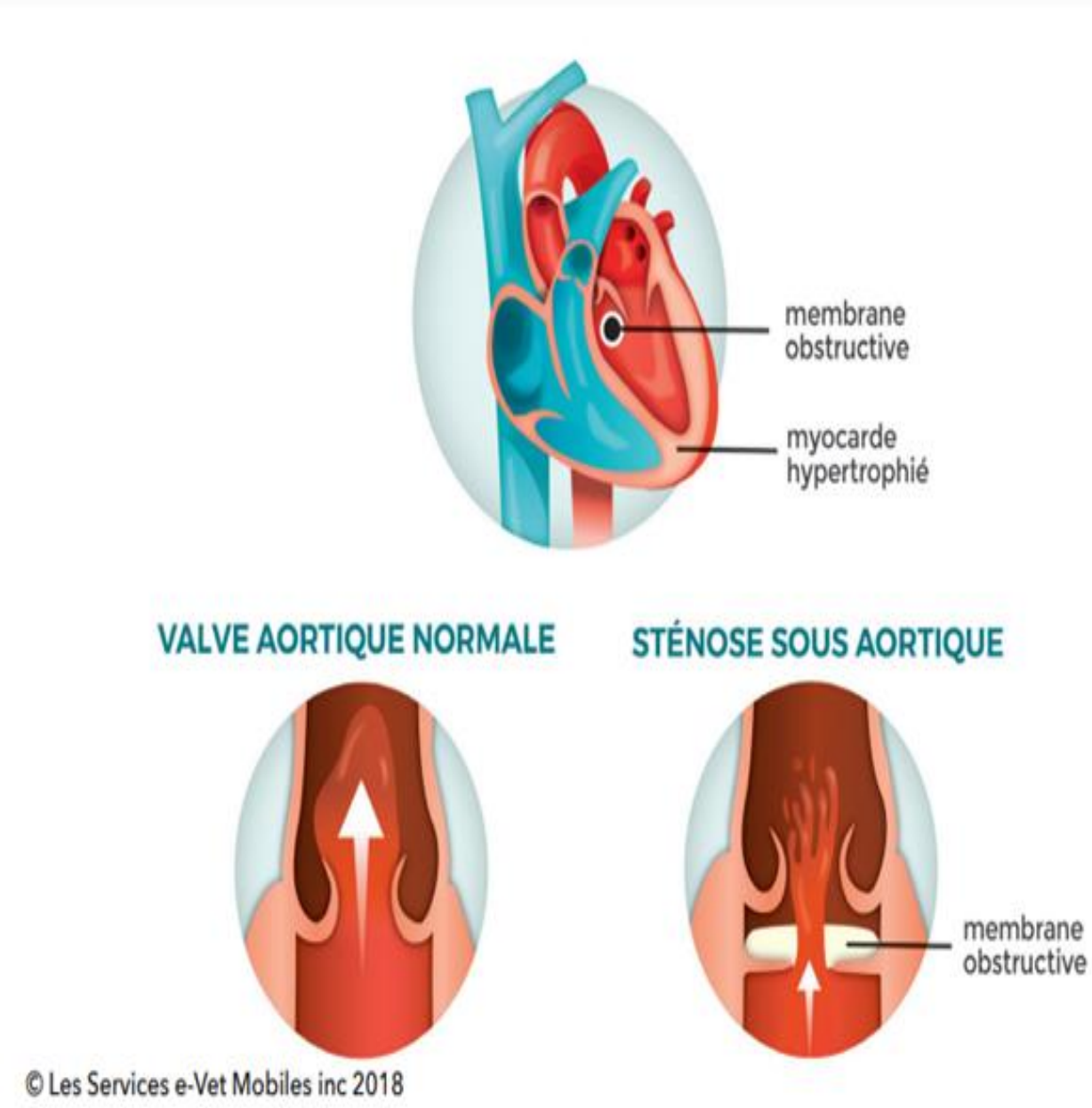
Dog breeds predisposed:

Newfoundland, Rottweiler, Golden Retriever, Boxer, Great Dane, German Shorthaired Pointer, German Shepherd, Belgian Tervuren, English Bulldog, Samoyed, Bull Terrier.

Pathophysiology

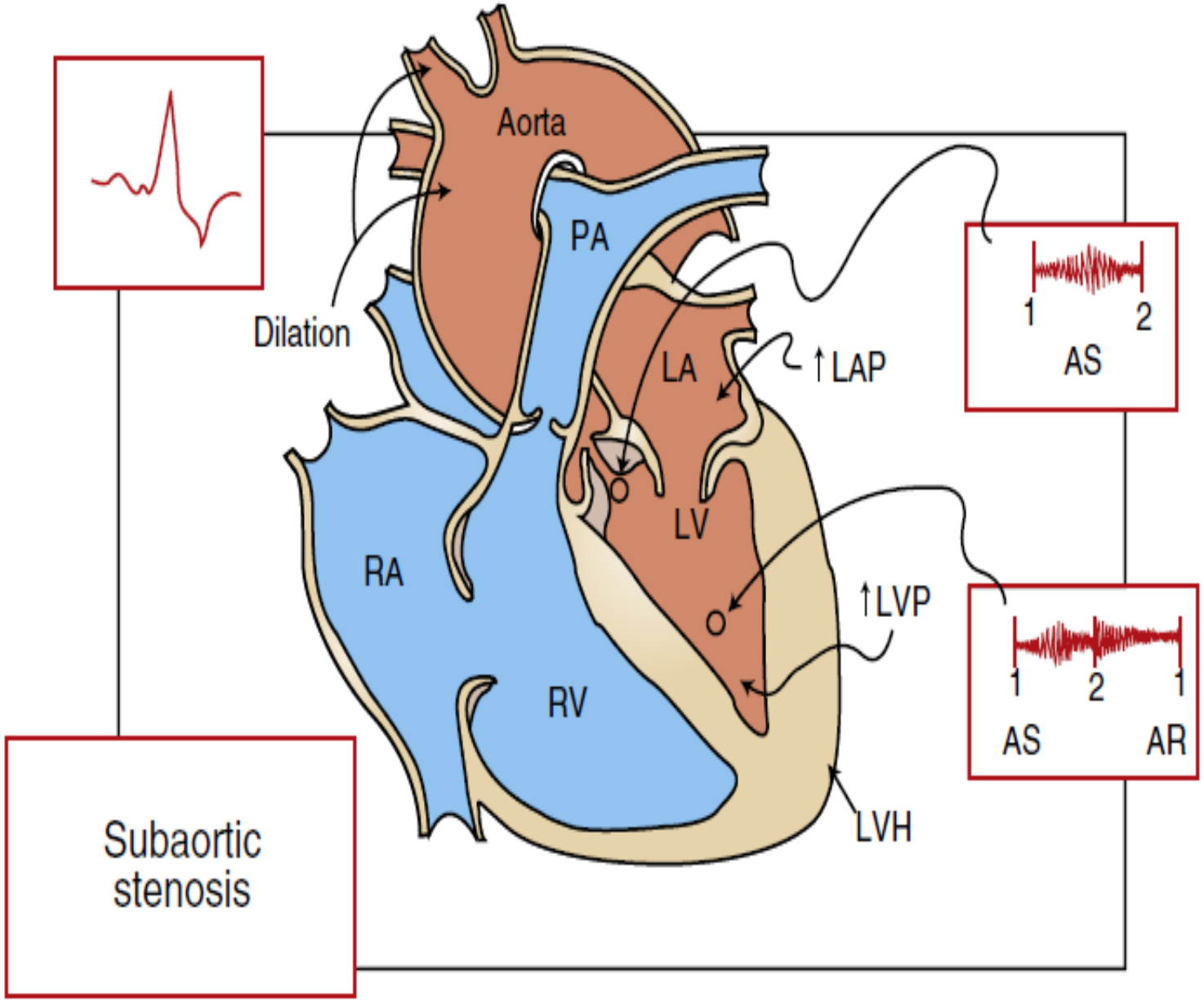
- Subvalvular stenosis results from the abnormal development of a fibrous (or fibromuscular) band, a fibrous ring, or a membranous tissue at the subvalvular level, which interferes with blood flow.
- The pressure overload induced on the left ventricle causes concentric hypertrophy of the left ventricle and, occasionally, left atrial enlargement.
- Turbulence and increased flow velocity through the stenotic outflow tract create a left-sided systolic basal murmur.

Aortic stenosis is a narrowing in the area of the aortic valve that obstructs blood flow from the left ventricle to the aorta.



- SAS predisposes to ventricular arrhythmias secondary to myocardial ischemia due to increased myocardial volume, decreased coronary blood flow, and myocardial fibrosis.
- Less frequently, increased pressure in the left atrium can lead to left-sided congestive heart failure (CHF).

Pathophysiology and genesis of clinical signs in congenital subaortic stenosis (SAS). LA, Left atrium; LAP, left arterial pressure; LV, left ventricle; LVH, left ventricular hypertrophy; LVP, left ventricular pressure; PA, pulmonary artery; RA, right atrium; RV, right ventricle.



Clinical presentation

- Animals with moderate to severe SAS often exhibit signs of growth retardation, exercise intolerance, and syncope.
- Tachypnea or dyspnea associated with the development of left-sided CHF occurs in older dogs.
- Animals that develop sustained rapid ventricular tachyarrhythmias may die suddenly.
- Physical examination reveals a left-sided basal systolic murmur whose degree correlates with the severity of SAS.
- Weak femoral pulses when stenosis is severe and, occasionally, arrhythmias.

Differential diagnosis

- Pulmonic stenosis (Pulmonic stenosis).
- Interventricular communication (CIV; Ventricular Septal Defect [VSD]).
- Physiological murmur.

Diagnosis

- ***Chest X-rays may show post-stenotic dilation of the aortic root.***

Aortic stenosis (lateral projection).
There is marked enlargement of the aortic arch cranial to the heart base (arrows).

- ***Echocardiography***

➢2D and M-mode

❑Allows visualization of the stenosis location and the fibrous anomaly.

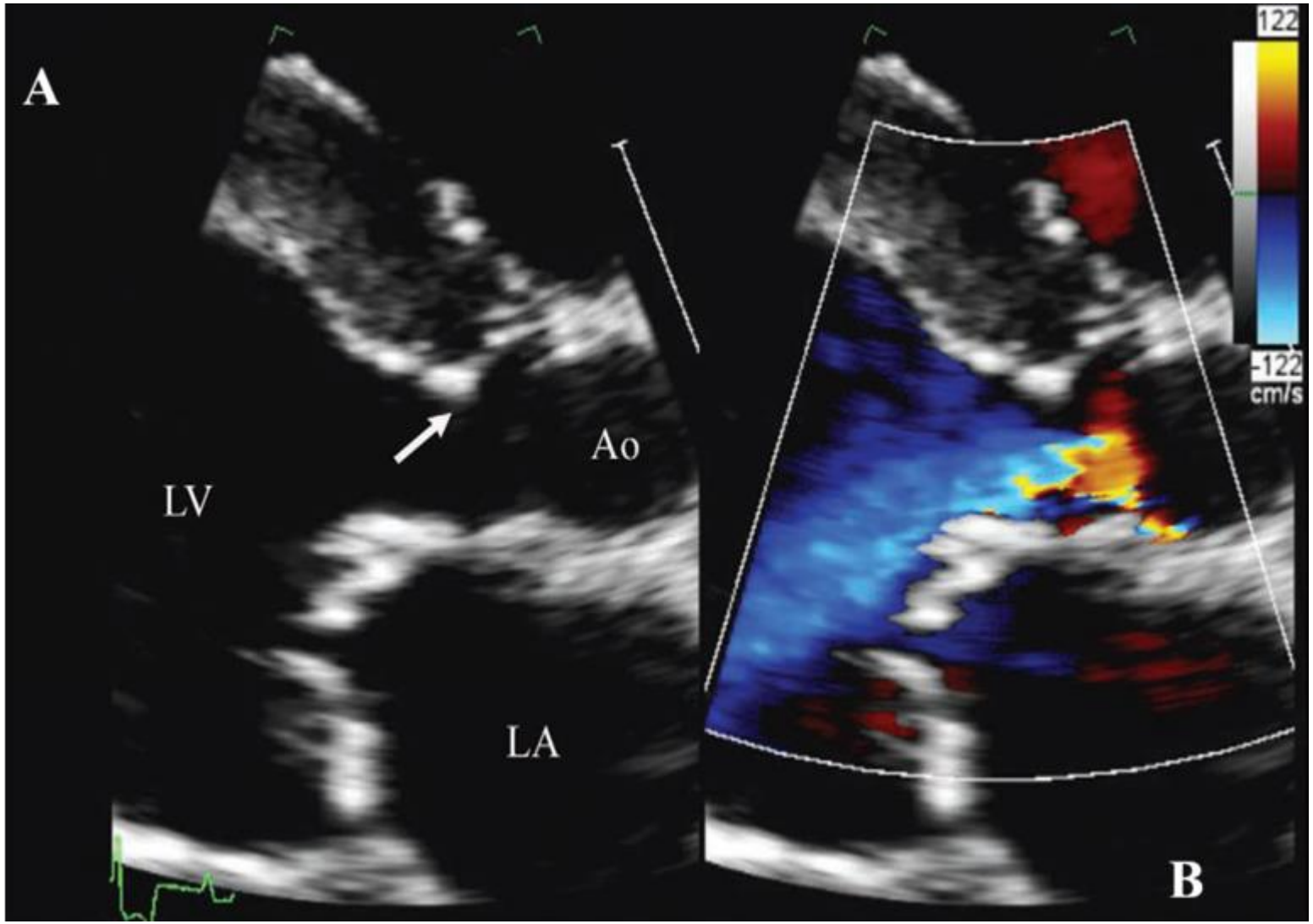
❑Concentric hypertrophy of the left ventricle in more severe cases.

❑Left atrial dilation in severe cases.

❑Post-stenotic dilation of the ascending aorta in moderate to severe cases.

- The color Doppler:
  - May show turbulent blood flow in the left ventricular outflow tract.
  - Mild aortic regurgitation is also usually present.
- Pulsed wave/continuous wave Doppler (determine the maximum systolic pressure gradient across the stenosis):
  - Most patients have velocities of 1.0 to 1.5 m/s, but up to 2.2 m/s are normal in some breeds.
  - Mild stenosis < 3.5 m/s (< 50 mmHg).
  - Moderate stenosis 3.5–4.5 m/s (50–80 mmHg).
  - Severe Stenosis > 4.5 m/s (> 80 mmHg).

(A) Right parasternal echocardiographic view in the long axis showing a sub-aortic ridge (arrow).
(B) The color Doppler reveals turbulence in the aorta beyond the stenosis = turbulent blood flow through the left ventricular outflow tract.



- For breeds with a high prevalence of SAS, programs that rely on auscultation and Doppler echocardiography to differentiate healthy dogs with physiological murmurs from animals with mild SAS have generally been developed.

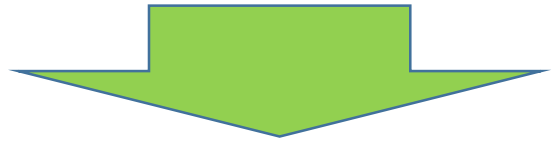
Treatment

- Cases of sub-aortic stenosis (regardless of the severity of the stenosis) require antibiotic therapy prior to any intervention (e.g., dental interventions) to reduce the risk of infectious endocarditis following bacteremia.
- Mild cases generally do not require any further treatment.

- In severe cases:

➢ ***Medical management:***

- ❑Due to the lack of effective interventional techniques, medical management serves as the foundation of treatment in most cases of SAS.
- ❑However, the benefits of medical treatment are largely theoretical and it has not been consistently demonstrated that they improve long-term outcomes.
- ❑Avoid extreme exertion, particularly sudden bouts of excitement or effort.
- ❑Ventricular arrhythmias controlled by antiarrhythmics (often beta-blockers: sotalol for example).
- ❑ACE inhibitors are sometimes prescribed to try to reduce myocardial remodeling or fibrosis.



- In cases of concentric left ventricular hypertrophy, use beta-blockers to improve diastolic filling and decrease myocardial oxygen consumption = they have no effect on survival but may improve clinical signs.
- Diuretic treatment is indicated in cases of CHF.
- Positive inotropes such as digoxin and pimobendan are contraindicated in SAS and other forms of flow obstruction and should not be used.

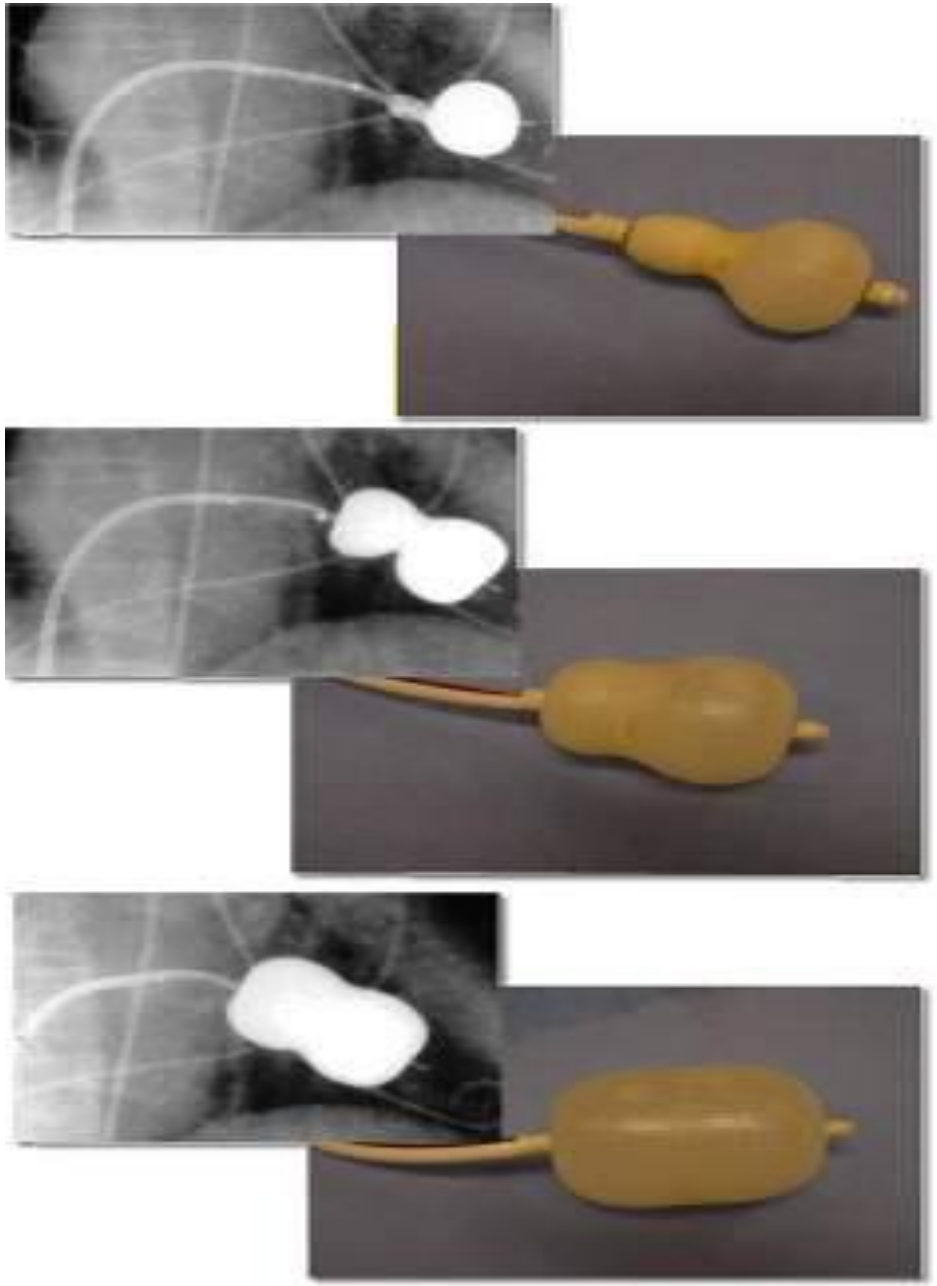
➢ ***Surgical resection.***

- Surgical resection of the lesion (fibrous tissue) of the SAS requires cardiopulmonary bypass (requires the establishment of extracorporeal circulation).
- Not indicated in patients with mild stenosis.
- Surgery in cases of severe SAS has not been associated with improved long-term survival and is rarely performed due to the cost and morbidity associated with the procedure.

➢ ***Balloon dilation***

- ❑ Balloon valvuloplasty is rarely performed and is most often considered in cases of discrete membrane stenosis.
- ❑ Special cutting and high-pressure balloons may be used.
- ❑ Significant reductions in the pressure gradient across the stenosis can be obtained immediately after balloon dilation.
- ❑ However, restenosis is likely and balloon dilation has not been systematically associated with improved long-term outcomes.

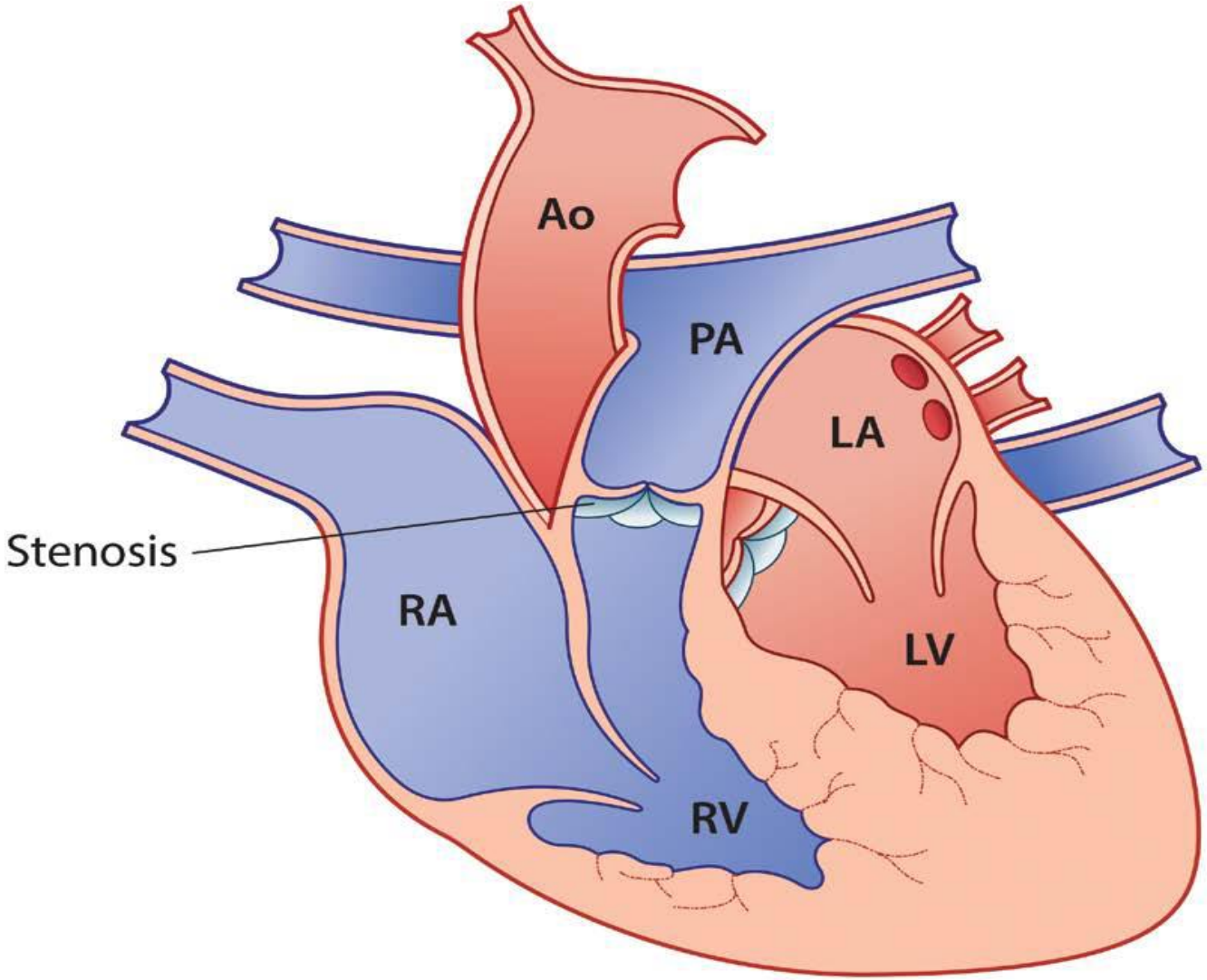
Balloon Dilation =
Balloon valvuloplasty.



Pulmonic Stenosis (PS)

Definition

- Pulmonary stenosis is among the most commonly reported congenital malformations in dogs.
- Pulmonary stenosis is less common in cats.
- It results from a narrowing of the right ventricular outflow tract in the area of the pulmonary valve and can occur at the subvalvular, valvular, or supravalvular levels.
- The most common manifestation in dogs is the narrowing at the valvular level.



Ao = Aorta
PA = Pulmonary artery
LA = Left atrium
LV = Left ventricle
RA = Right atrium
RV = Right ventricle
PDA = Patent ductus arteriosus

Pulmonary Stenosis (Pulmonic Stenosis [PS])

- The breeds predisposed to pulmonary stenosis (PS) include Boxers, Beagles, Cocker Spaniels, Miniature Schnauzers, English Bulldogs, and terriers.

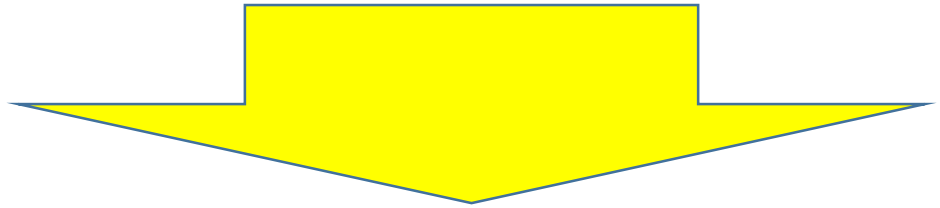
Beagle

Etiology

• Pulmonary Stenosis is generally valvular in dogs; two forms are observed:

- ❑Type A: normal (thin) pulmonary valve leaflets but partially fused.
- ❑Type B: hypoplastic valvular ring with thickening of the valve leaflets.
- ❑Some patients have a mixed type, with components of type A and type B.

- Pulmonary stenosis can be mild to severe =

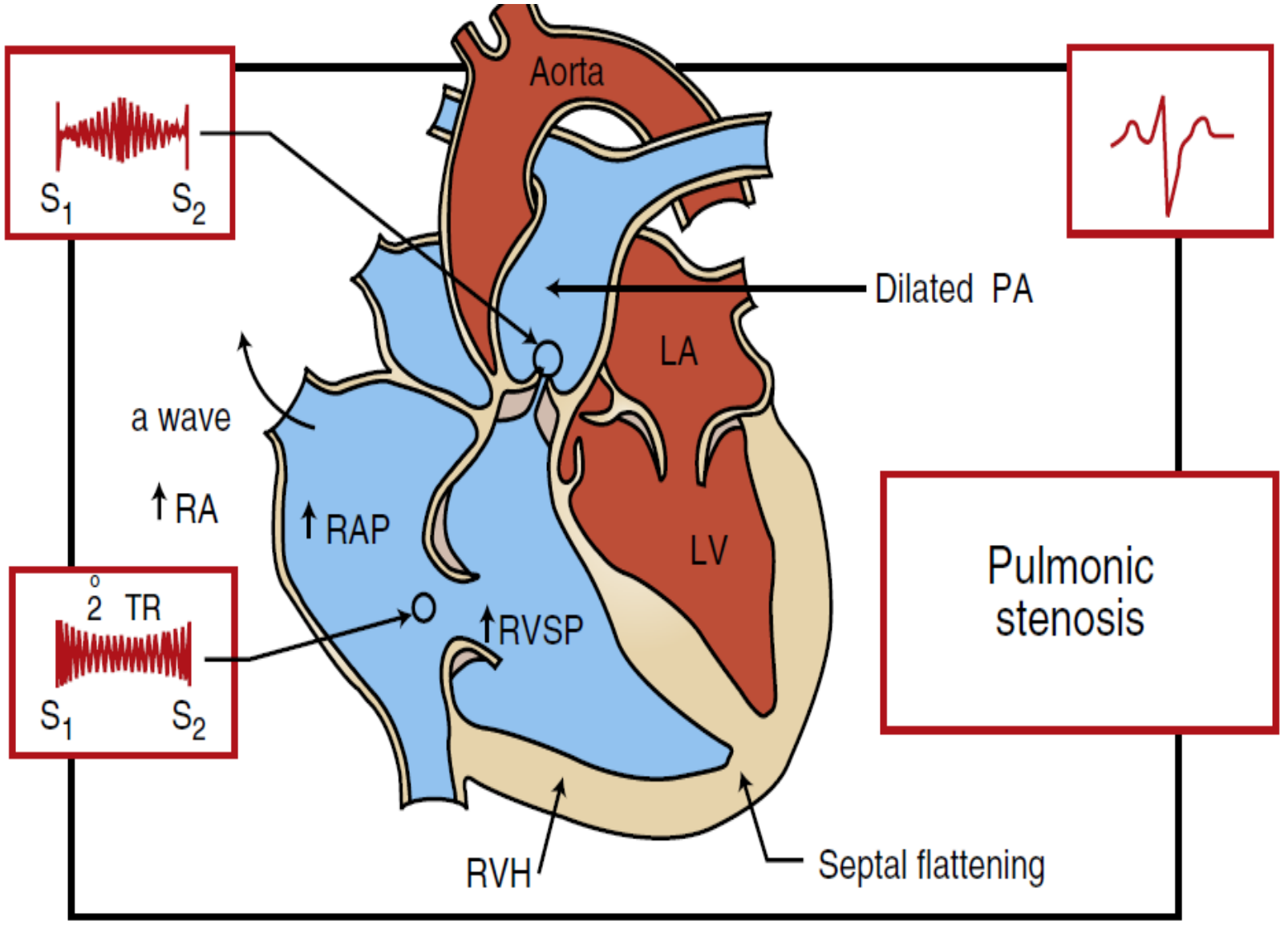


In very severe stenoses, increased afterload leads to right ventricular hypertrophy and may eventually lead to right atrial dilation and right heart failure as well as ventricular arrhythmias.

Pathophysiology

- Blood flow obstruction leads to increased pressure in the right ventricle, causing concentric hypertrophy of the right ventricle and myocardial fibrosis with possible dilation of the right atrium.
- Turbulent blood flow and increased velocity through the stenotic valve lead to a left basal systolic murmur.
- Patients with severe pulmonary stenosis develop right diastolic dysfunction and are at risk of eventual right heart failure due to chronic pressure overload.
- Tricuspid valve dysplasia (TVD) is an occasional concomitant congenital defect and leads to exacerbation of right heart dysfunction and more severe clinical signs.

Pathophysiology and genesis of clinical signs in pulmonic stenosis (PS). LA, Left atrium; LV, left ventricle; PA, pulmonary artery; RAP, right atrial l pressure; RVH, right ventricular hypertrophy; RVSP, right ventricular systolic pressure.



Clinical presentation

- Most animals affected by PS show no clinical signs or present mild clinical signs.
- About one-third of dogs with PS who present clinical signs often show dyspnea, lethargy, exercise intolerance, syncope, sudden death, and growth retardation.
- In later stages (related to the severity of the stenosis), signs associated with right heart failure may be present.

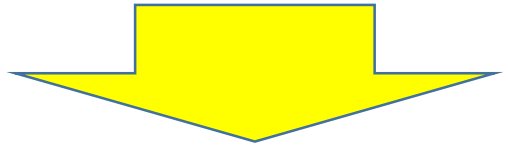
- The clinical examination may reveal the following:

❑Precordial shock (apical shock, Point of Maximum Intensity [PMI]) may be stronger on the right due to right ventricular hypertrophy.

❑Left basal systolic murmur crescendo-decrescendo (sometimes with a concurrent right apical murmur in case of tricuspid regurgitation).

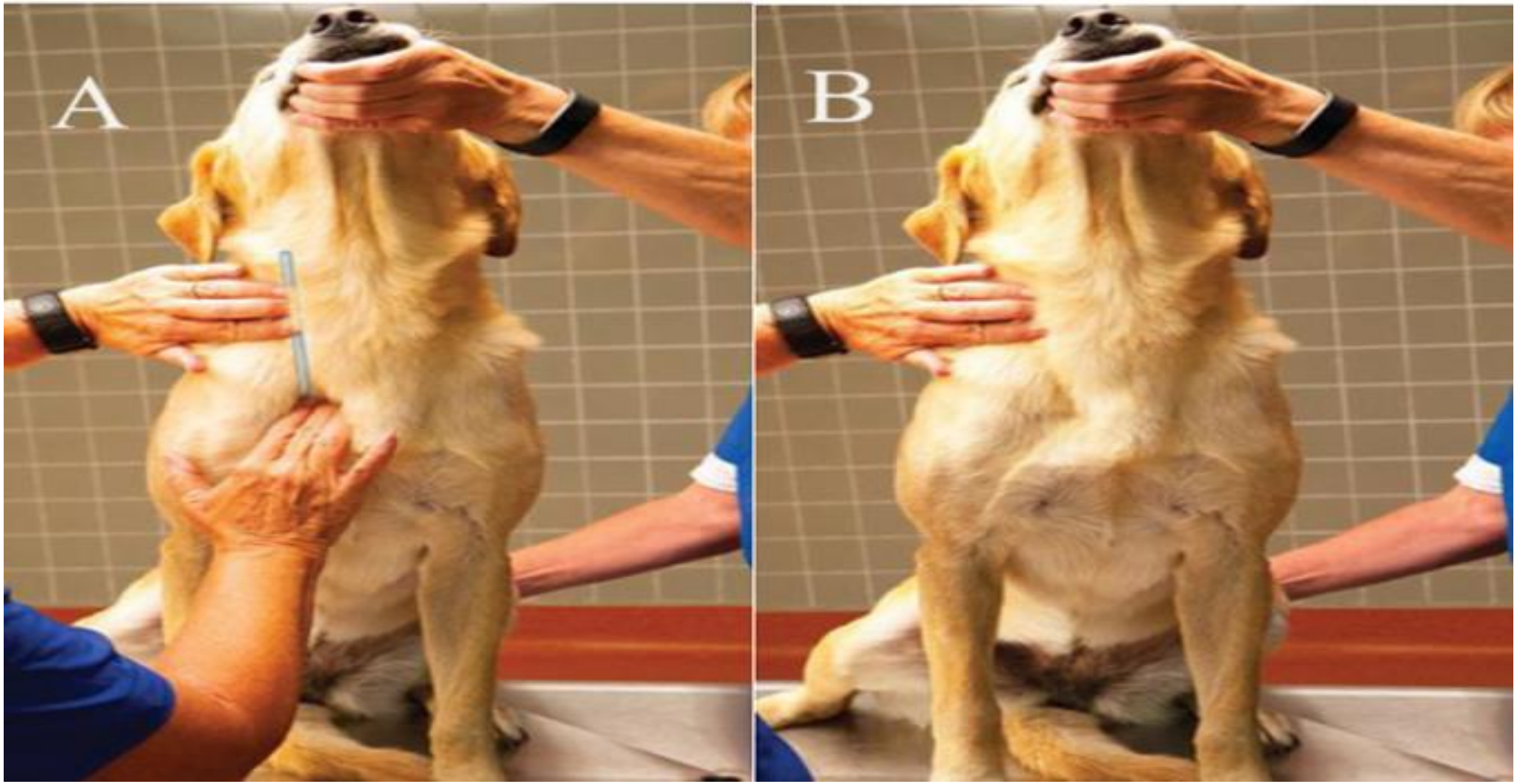
❑Degree of murmur related to the severity of the stenosis.

❑Normal arterial pulse (Good pulse quality).



Palpation of the precordial impulse in dogs.

- Prominent jugular pulse: Jugular distension is associated with increased right atrial pressure.
- In cases of right heart failure, abdominal palpation may reveal the presence of ascites and hepatomegaly.
- The presence of a patent foramen ovale at the atrial level may act as a site for a right-to-left shunt, resulting in cyanosis, hypoxemia, and polycythemia.



Venous pulse = A/Palpation and digital obstruction of the jugular vein; B/ Once it is no longer obstructed, the jugular vein should not be distended.

Severe ascites in a dog with right heart failure caused by severe pulmonary stenosis and severe tricuspid dysplasia.

Differential diagnosis

- Subvalvular aortic stenosis (Subvalvular aortic stenosis [SAS]).
- Tetralogy of Fallot (Tetralogy of Fallot [ToF]).
- Ventricular septal defect (CIV; Ventricular Septal Defect [VSD]).
- Physiologic murmur (physiologic murmur).

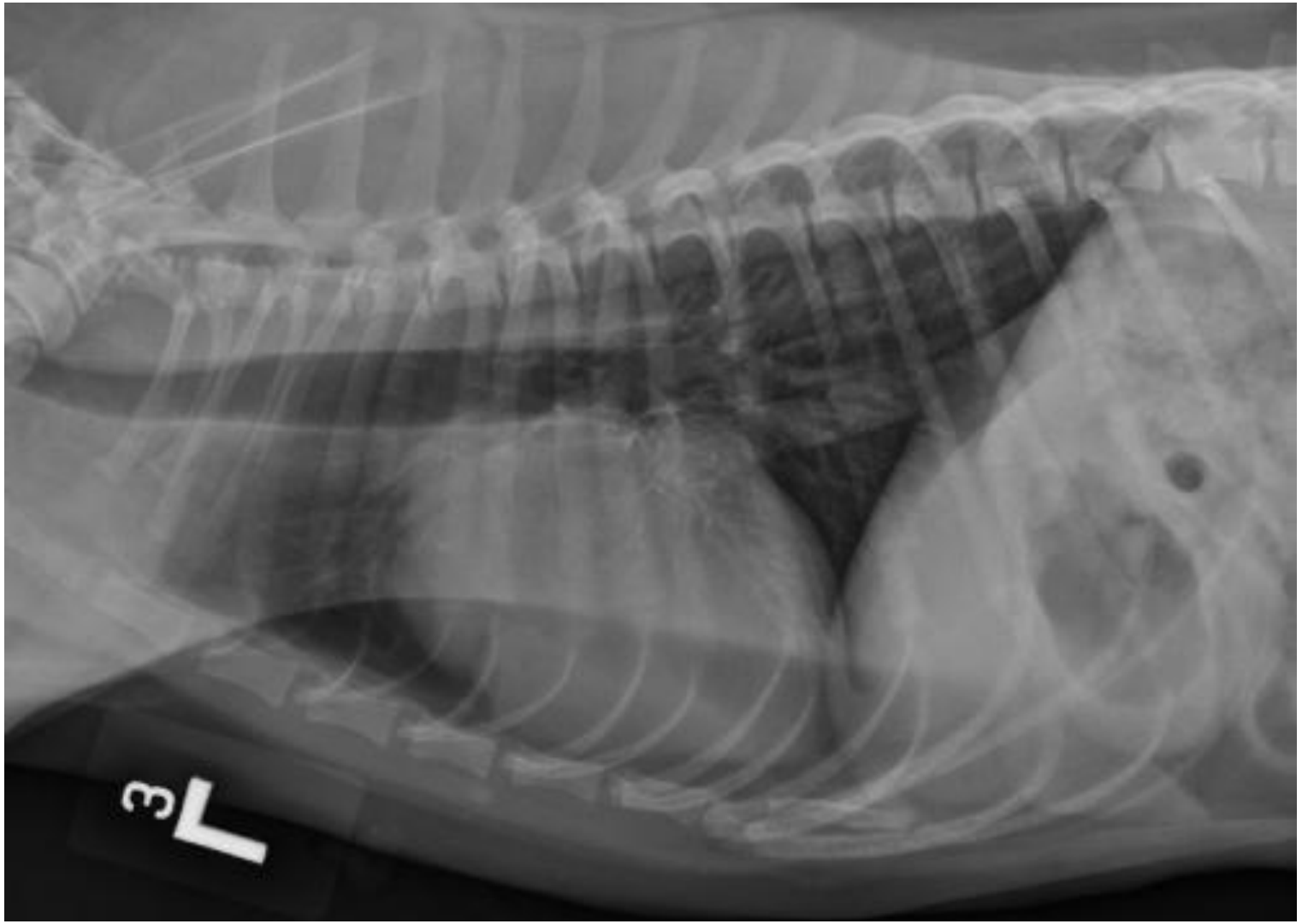
Diagnosis

- ***Chest X-rays***

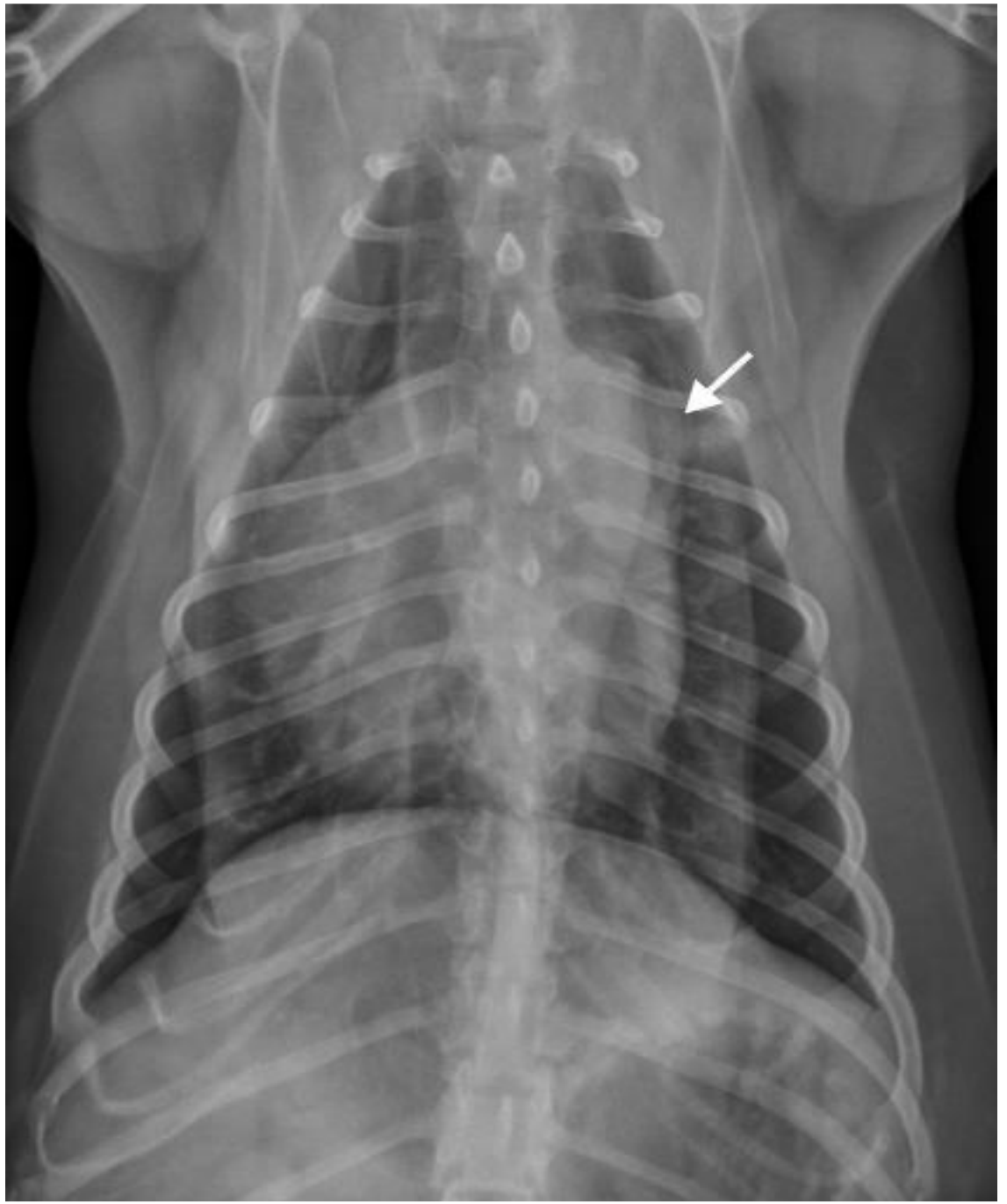
Chest X-rays often show the presence of:

- ❑Post-stenotic dilatation of the pulmonary trunk.
- ❑Right cardiomegaly.

Pulmonary stenosis (PS) (left lateral view) in a dog. There is increased sternal contact on the right side of the heart due to hypertrophy on the right side of the heart and normal pulmonary vascularization.

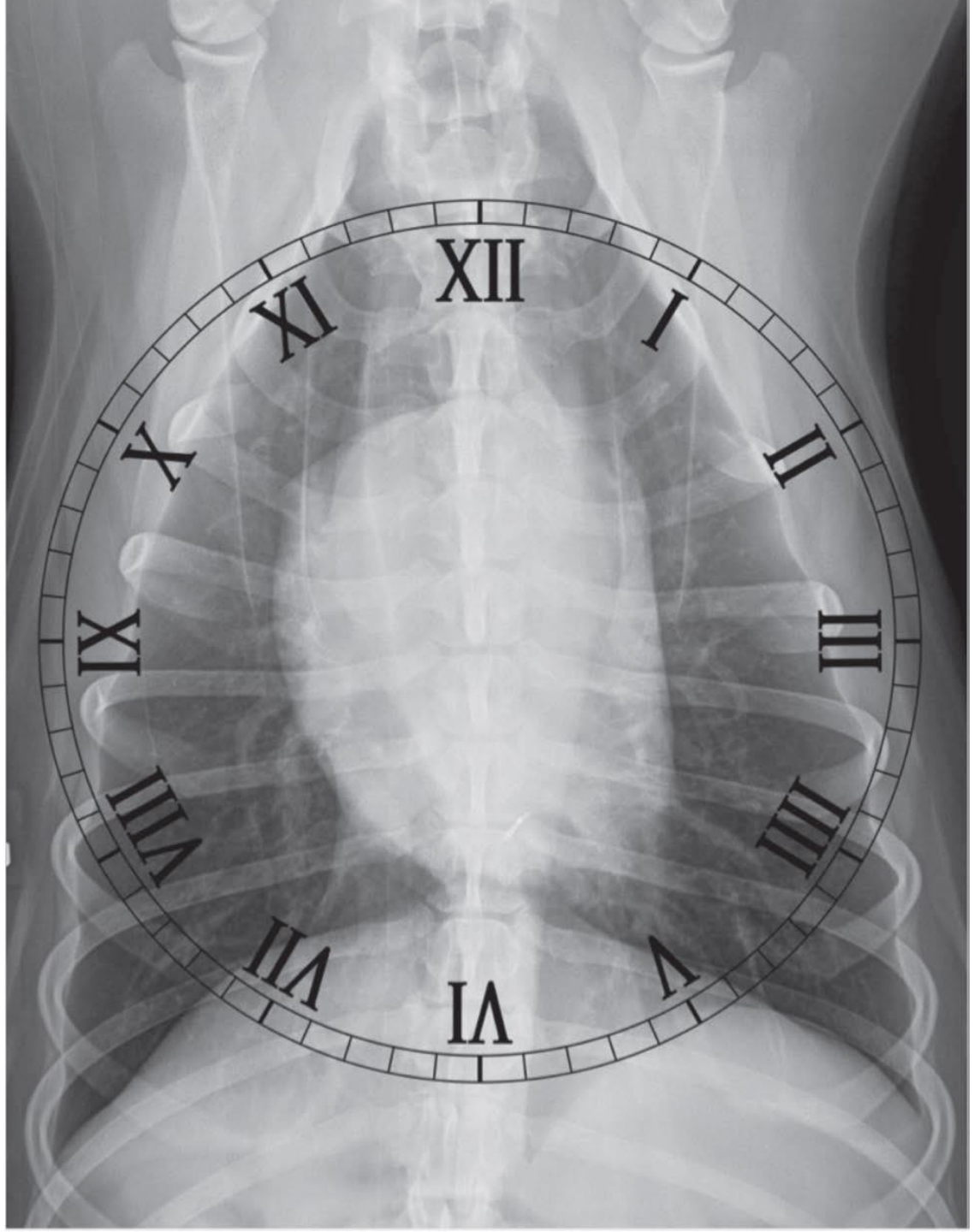


Pulmonary stenosis (PS) (dorso-ventral view) in a dog. There is marked cardiomegaly of the right heart and prominence of the main pulmonary artery (arrow).

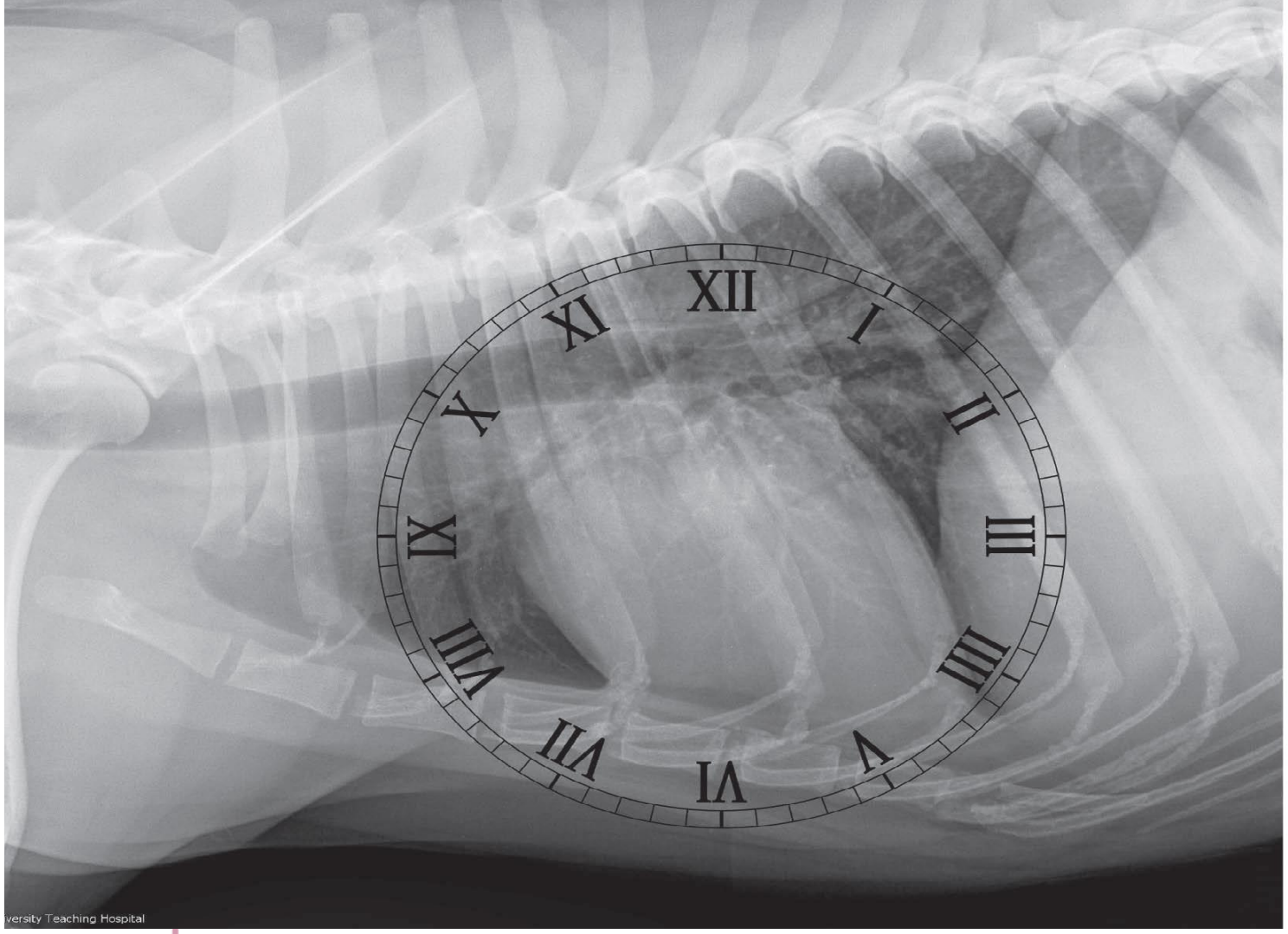


Position of the heart chambers using the clock dial analogy (Dorso-ventral incidence).

Time:	Cardiac chamber:
11:00 to 1:00	Aorta
1:00 to 2:00	Main pulmonary artery
2:00 to 3:00	Left auricle
3:00 to 5:00	Left ventricle
5:00 to 9:00	Right ventricle
9:00 to 11:00	Right atrium
Center dial (between mainstem bronchi)	Left atrium



Position of the cardiac cavities using the analogy of the clock face (Lateral incidence).



Time:	Cardiac chamber(s):
12:00 to 3:00	Left atrium
3:00 to 5:00	Left ventricle
5:00 to 8:00	Right ventricle
8:00 to 11:00	Right atrium, main pulmonary artery, ascending aorta and arch

- ***Echocardiography***

➢2D echocardiography and M-mode can show:

❑No obvious abnormalities are visible in mild cases.

❑Concentric hypertrophy of the right ventricle in the most severe cases.

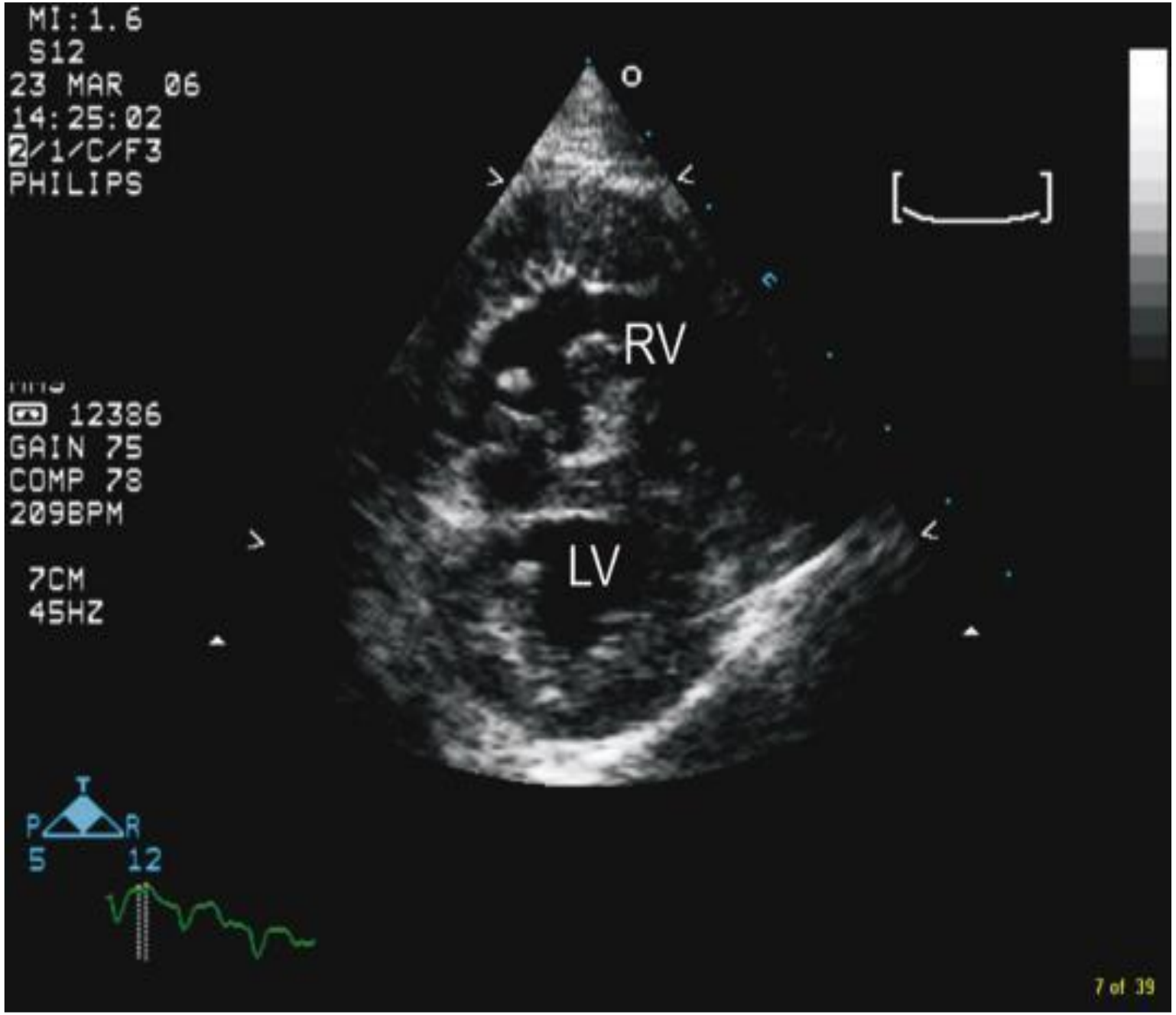
❑Fusion of the pulmonary valve leaflets and/or hypoplasia of the pulmonary artery with abnormal pulmonary valves.

❑Post-stenotic dilation of the pulmonary artery in moderate to severe cases.

❑Right atrial dilation in severe cases.

❑Due to increased pressures in the right ventricle, the septum is usually flattened.

Right parasternal short-axis, two-dimensional (2D) echocardiogram obtained from a West Highland White Terrier with severe pulmonic stenosis (PS). Note the severely thickened right ventricle (RV). LV, Left ventricle.



- Pulsed wave/continuous wave Doppler offers a sensitive and non-invasive method to determine the severity of stenosis by estimating the maximum systolic pressure gradient across the stenosis.:

- Most patients have speeds of about 1.0 m/s.
- Mild stenosis < 3.5 m/s (< 50 mmHg).
- Moderate 3.5–4.5 m/s (50–80 mmHg).
- Severe > 4.5 m/s (> 80 mmHg).

- ***Electrocardiography***

Signs of right ventricular hypertrophy are present in the majority of cases:

- ❑Deep S waves in leads I, II, III, and aVF.
- ❑Right axis deviation.
- ❑Ventricular arrhythmias may be present.

Prognosis

- The prognosis varies depending on the severity of the stenosis:

❑Mild to moderate disease may have no effect on life expectancy.

❑Patients with severe stenosis (who do not undergo intervention or who present severe stenosis after an intervention) have an increased risk of developing arrhythmias, heart failure, and sudden death.

❑Severe cases: They depend on the results of balloon valvuloplasty (or another surgical intervention).

Treatment

- The goal of treatment is to reduce the overload of right ventricular systolic pressure:

❑Mild cases do not require treatment.

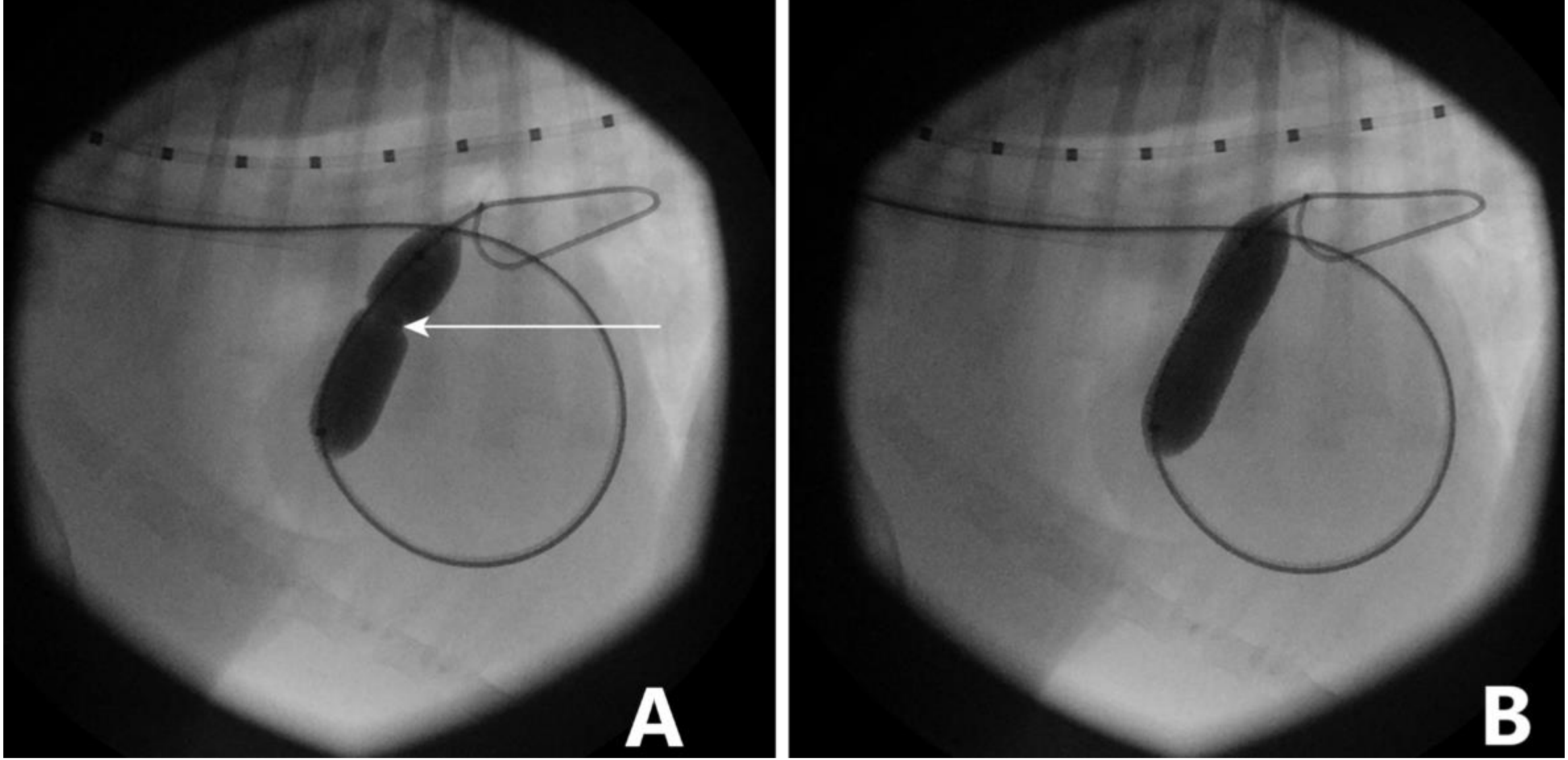
❑In cases of mild PS or mild clinical signs, beta-blockers (atenolol 0.25 -1.5 mg/kg PO q12h) may be used to reduce the myocardial oxygen demand (negative inotropes) and for their antiarrhythmic properties.

❑In cases of moderate to severe PS, balloon valvuloplasty or surgical procedures are recommended.

- Balloon valvuloplasty:

❑This is a particularly effective technique for patients with thin and fused pulmonary valve leaflets.

❑Success is considered with a 40 to 50% reduction in pressure gradient.

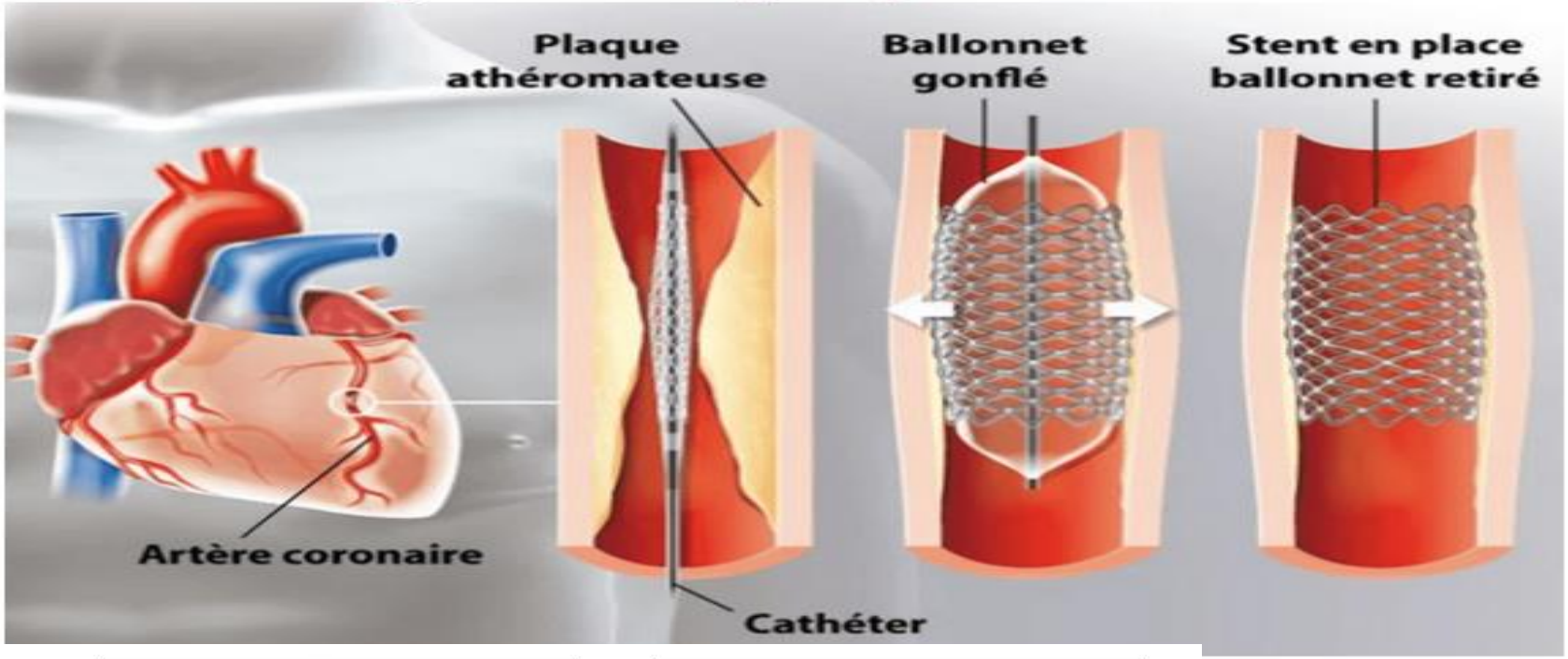


Fluoroscopy image showing a balloon catheter across the pulmonary valve. (A) The arrow indicates the stenosis. And (B) fluoroscopy image showing that the stenosis has disappeared.

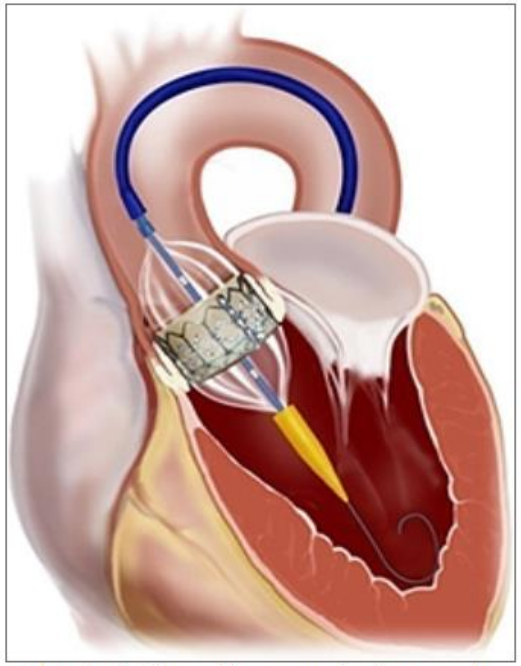
Alternatives exist for patients for whom balloon valvuloplasty is not considered appropriate (e.g., hypoplastic pulmonary arteries):

- Pulmonary stent: Limited availability.
- Surgical options: In general, surgical correction is not commonly practiced (limited availability)

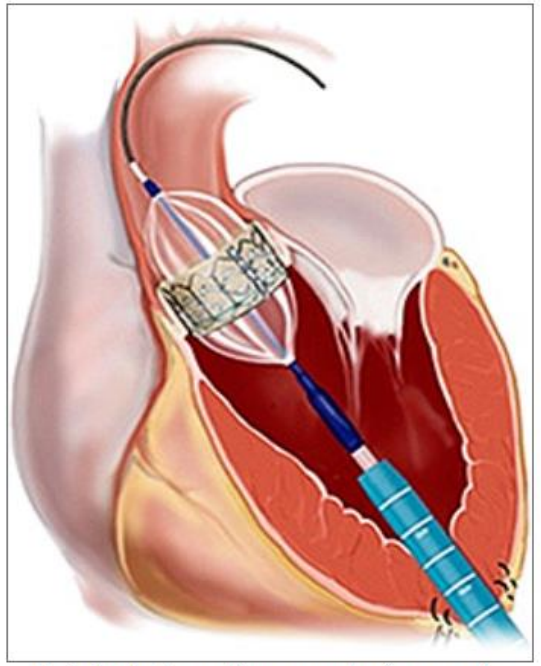
Principle of stent placement



Bioprothèse Edwards SAPIEN®



Valvuloplastie aortique percutanée par voie rétrograde ou transfémorale



Valvuloplastie aortique percutanée par voie antérograde ou transapicale

Tetralogy of Fallot (ToF)

Definition

- Tetralogy of Fallot is a cyanotic congenital heart defect comprising the following 4 anomalies:

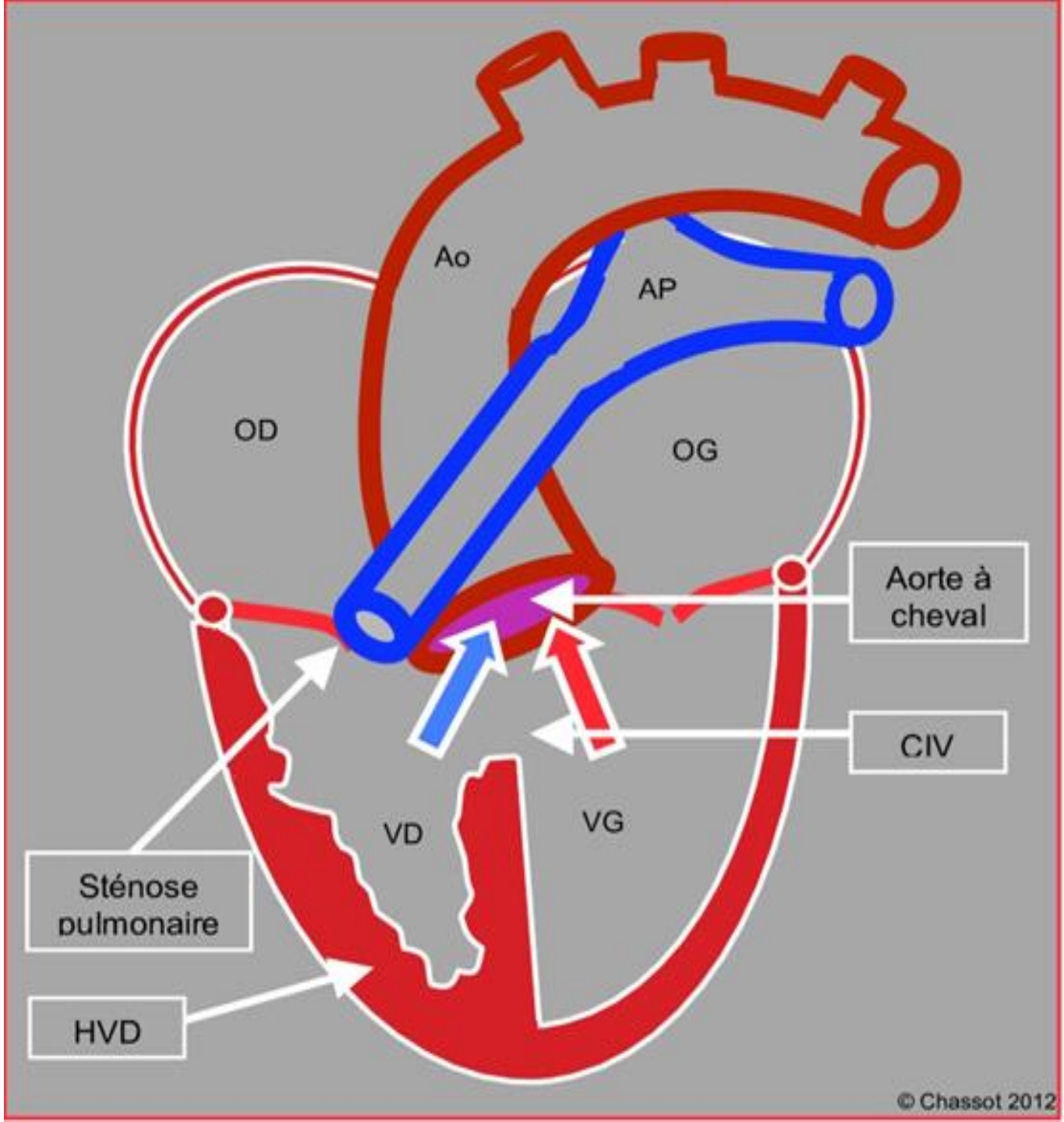
❑ Ventricular septal defect (VSD).

❑ Overriding aorta (dextroposition of the aorta; Overriding aorta).

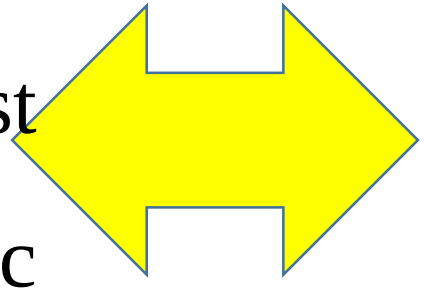
❑ Pulmonary stenosis (Pulmonic Stenosis [PS]).

❑ Right ventricular hypertrophy.

Illustration of Tetralogy of Fallot which includes 4 distinct anomalies: The aortic valve straddles a ventricular septal defect (VSD). The valve and the pulmonary artery are stenosed. Right ventricular hypertrophy (RVH). The aorta receives a mixture of oxygenated blood from the left ventricle and deoxygenated blood from the right ventricle through the VSD.



Tetralogy of Fallot is a rare heart condition in dogs and cats, yet it represents the most frequent cyanotic congenital anomaly.



Blood tends to flow from right to left, mixing deoxygenated blood in the systemic circulation; erythrocytosis (polycythemia) secondary to hypoxemia.

- Breeds predisposed to Tetralogy of Fallot include the English Bulldog, the Keeshond, the Miniature Poodle, the Miniature Schnauzer, and the Wire-Haired Fox Terrier.
- This defect has also been recognized in other dog breeds and in cats.

Wolf-Spitz (Keeshond)

Wire-haired Fox Terrier (Wire-Haired Fox Terrier)

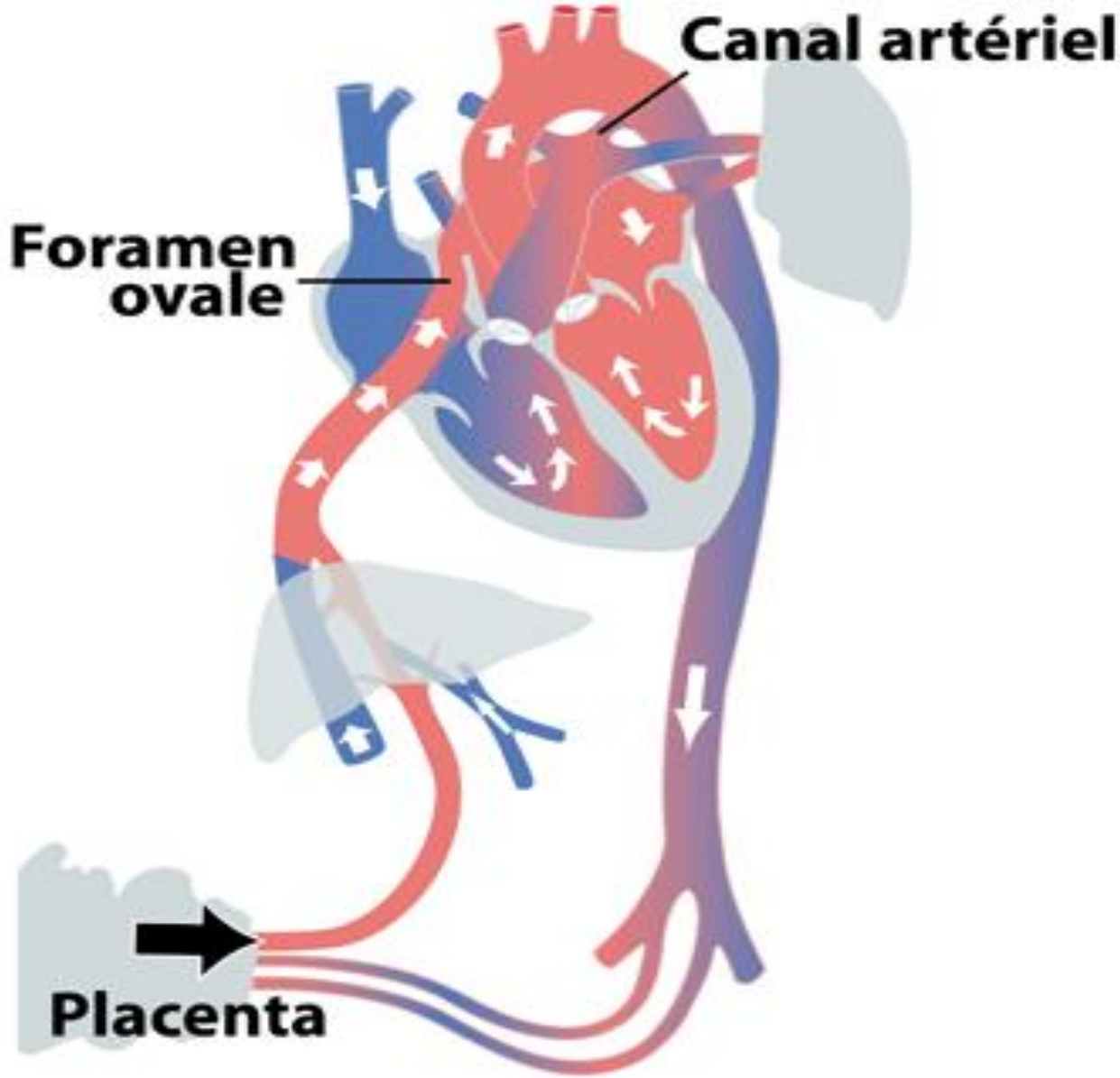
Etiology

- Congenital anomaly of the division of the fetal arterial duct during the formation of the aorta and the pulmonary artery.
- Unequal division of the fetal arterial duct results in:

❑A wide aorta and a small pulmonary artery, leading to an inability of the division septum to fuse with the interventricular septum.

❑This creates a ventricular septal defect (VSD) and leaves the right ventricle exposed to systemic pressures.

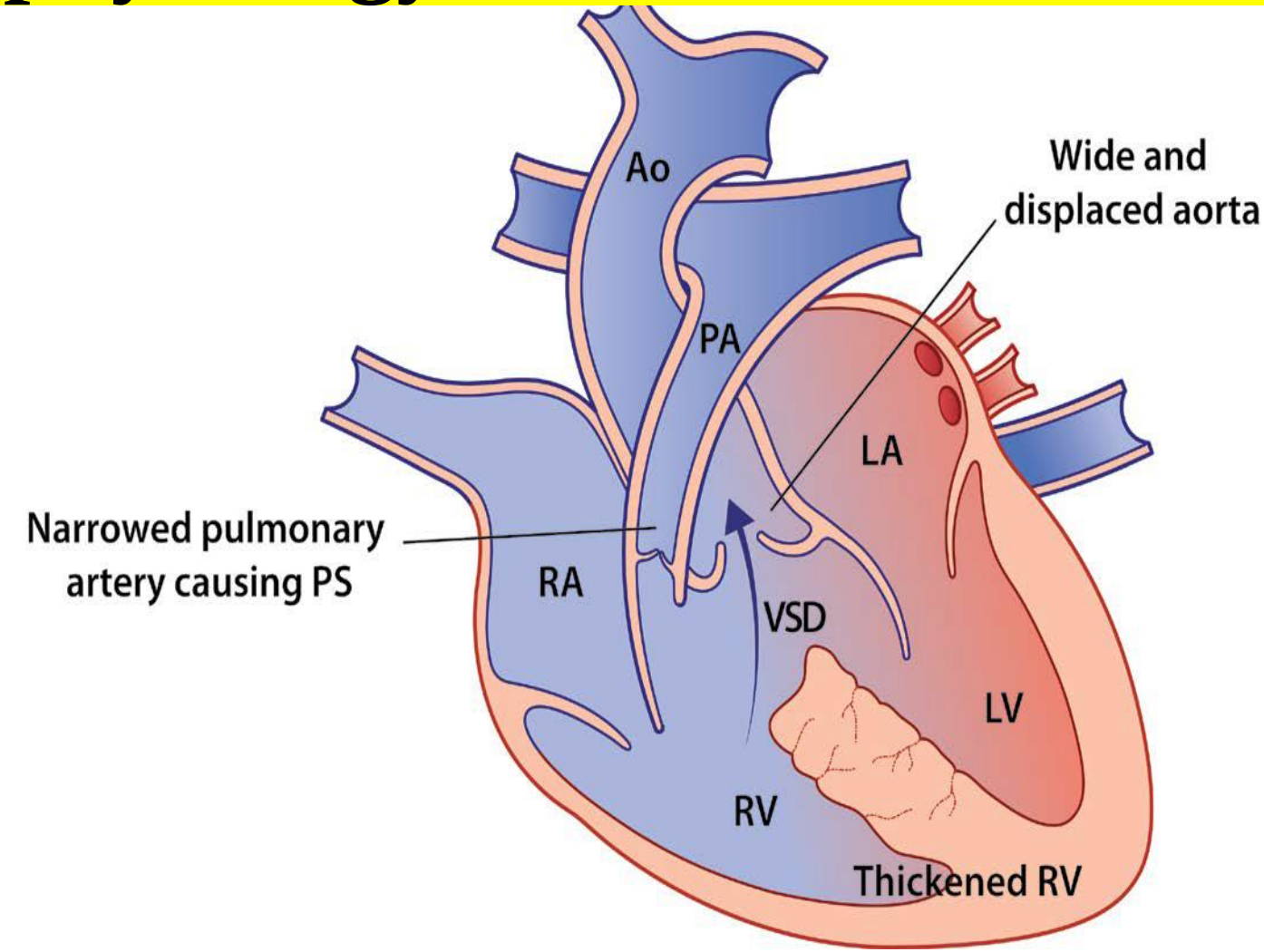
- Right ventricular hypertrophy is the result of the high pressure in the right ventricle due to pulmonary hypoplasia (pulmonary stenosis).


Canal artériel
Foramen ovale
Placenta

Pathophysiology

Tetralogy of Fallot includes the following 4 defects:

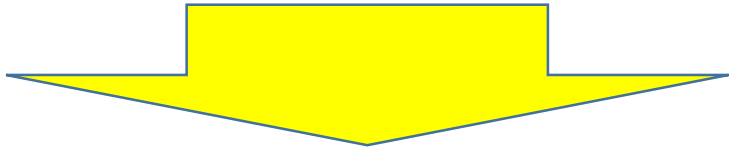
- Pulmonary stenosis (PS): The pulmonary artery is either narrowed or hypoplastic (underdeveloped).
- Overriding aorta: The aorta is large and displaced.
- Large ventricular septal defect (VSD): Due to poor alignment of the interventricular septum.
- Right ventricular hypertrophy: Increased pressure on the right ventricle due to pulmonary stenosis.



Tetralogy of Fallot. Note how deoxygenated and oxygenated blood mixes in the aorta due to the VSD.

Due to the high pressure in the right ventricle caused by PS, blood flows freely through the VSD to the left ventricle (right-to-left shunt).

- The oxygenated blood then mixes with deoxygenated blood, which is then ejected into the systemic circulation via the aorta.
- This causes hypoxemia, which the body attempts to compensate for by increasing red blood cell production, resulting in marked polycythemia.



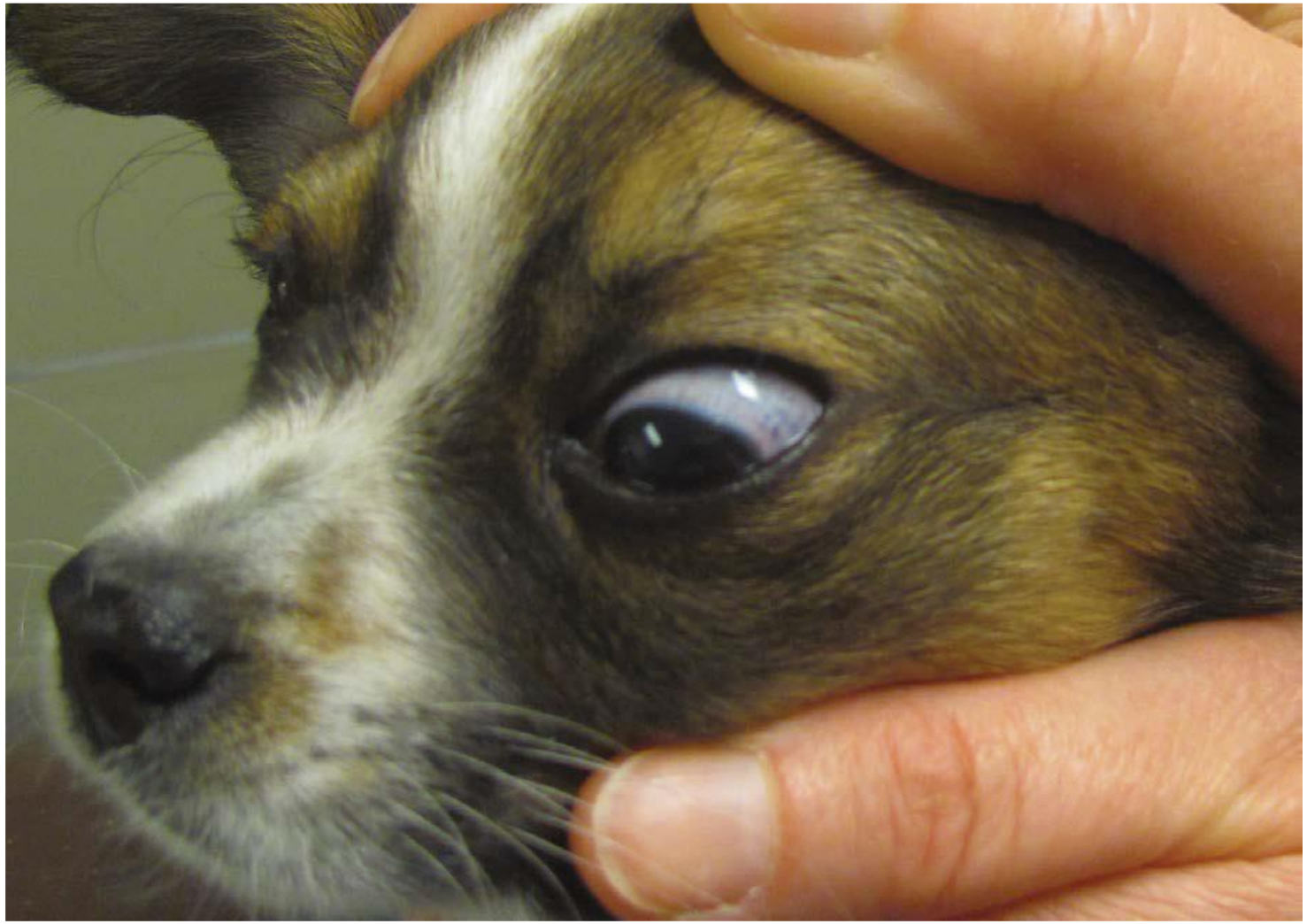
Consequences = Cyanosis + hypoxemia + polycythemia.

Clinical presentation

- The severity of clinical signs varies according to the degree of abnormalities.
- Main signs/ patients generally present with:

- ❑ Mucosal cyanosis.
- ❑ Shortness of breath during periods of exertion/excitement.
- ❑ Severe exercise intolerance.
- ❑ Growth delay (with poor health from an early age).
- ❑ Syncope.

Mildly cyanotic tongue in a 2-year-old male Collie with tetralogy of Fallot.



Cyanosis is apparent in the sclera of this 2-month-old, mixed-breed dog with tetralogy of Fallot.

- Animals affected by tetralogy of Fallot may develop episodes of breathing difficulties when the right-to-left shunt worsens, causing systemic hypoxemia.
- Tetralogy of Fallot is usually associated with a systolic murmur from the PS and/or the VSD.
- Potential developments:

❑ Right congestive heart failure (CHF): Rare outcome.

❑ Seizures/obnubilation due to polycythemia.

❑ Sudden death.

The clinical examination reveals the following:

- The mucous membranes are more obviously cyanotic after exercise/excitement due to the drop in peripheral vascular resistance and the worsening of the right-to-left shunt.
- Heart murmur:
  - Pulmonary stenosis is often the most obvious component of the murmur: left basal systolic murmur crescendo-decrescendo.
  - A right apical systolic murmur due to tricuspid regurgitation may also be heard.
- The quality of the pulse is generally normal.

Differential diagnosis

- Interventricular communication (IVC; Ventricular Septal Defect [VSD]).
- Pulmonary stenosis (Pulmonic Stenosis [PS]).

Diagnosis

- Laboratory results: Erythrocytosis/polycythemia.
- Electrocardiography:
  - ❑ Deep S waves in leads I, II, III, and aVF.
  - ❑ Rightward deviation of the cardiac electrical axis.
- Chest X-rays show cardiomegaly of the right heart and pulmonary under-circulation.

- Echocardiography allows:

❑To visualize the VSD.

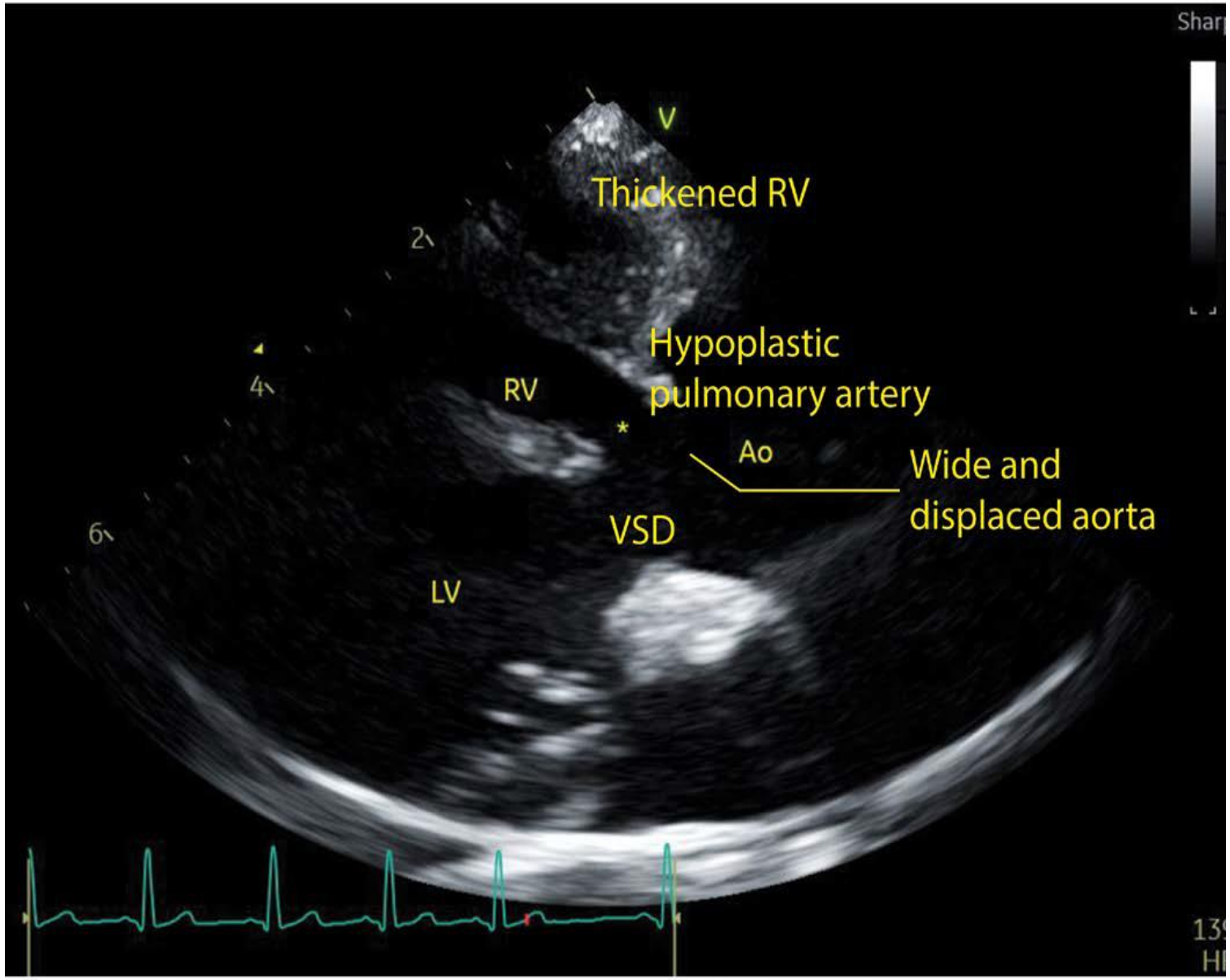
❑To assess the severity of PS.

❑To visualize the displacement of the aortic root to the right side.

❑To visualize right ventricular hypertrophy.

❑A color flow Doppler is necessary to visualize the shunt.

Tetralogy of Fallot visible on echocardiography. Note right ventricular hypertrophy (shown), the VSD allowing communication between the left and right ventricles, and the wide aorta (shown). RV = Right ventricle. Ao = Aorta. LV = Left ventricle. VSD = ventricular septal defect.



Management

- Medical management:

❑ Avoid agents that cause systemic vasodilation, such as acepromazine.

❑ Limit right-to-left shunting and hypoxemia by administering non-selective beta-blockers orally (propranolol).

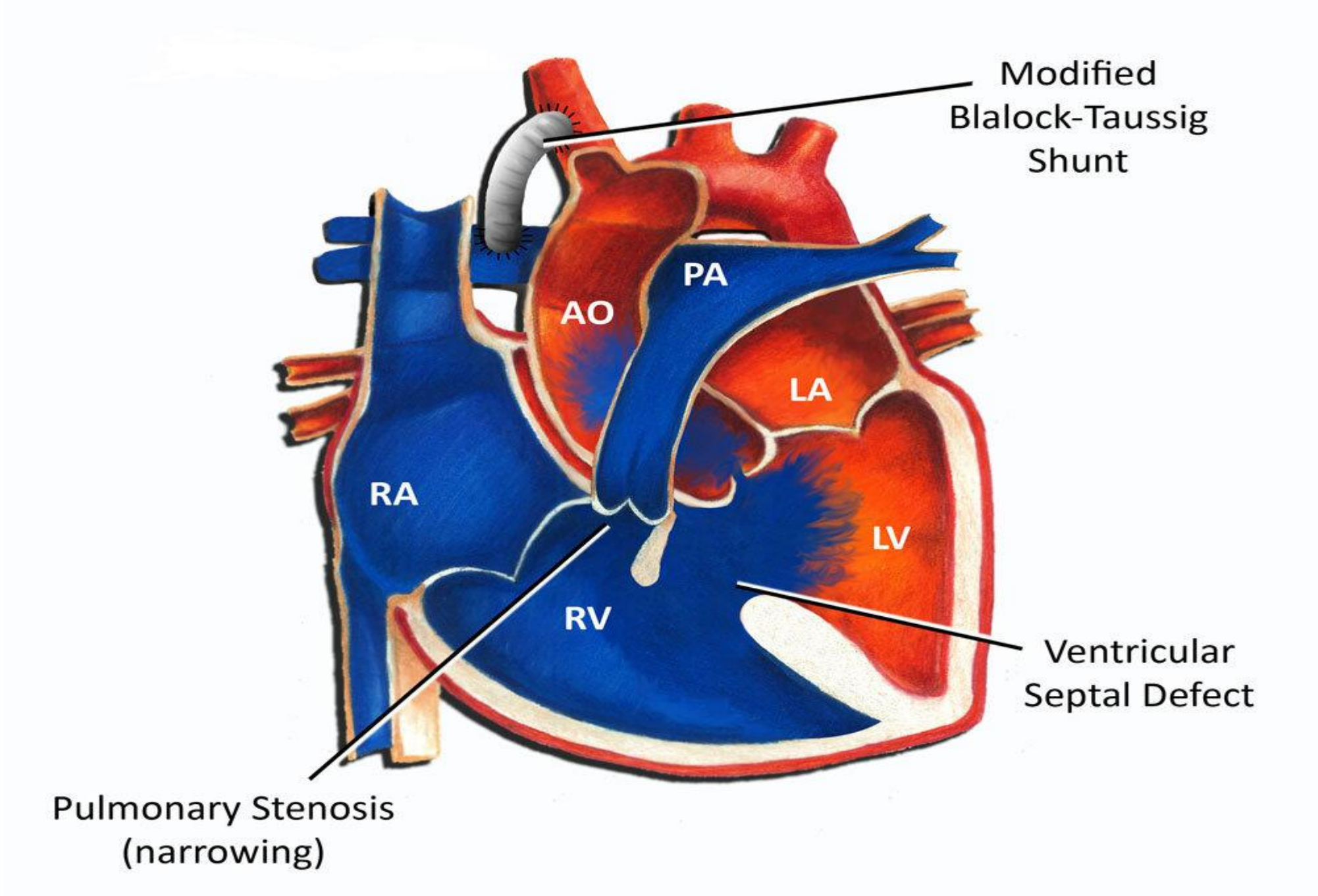
- Phlebotomies (to maintain hematocrit at 55–65 %) are indicated when polycythemia is responsible for clinical signs (usually for a hematocrit [packed cell volume (PCV)] >70 %).

In case of acute crisis, it is recommended:

- Complete rest.
- Intravenous fluid therapy.
- Oxygen therapy.
- Sodium bicarbonate in case of acidosis.

- Surgical management:

- ❑Closure of the interventricular communication; requires cardiopulmonary bypass (rarely performed).
- ❑Creation of a subclavian artery-pulmonary artery shunt (modified Blalock-Thomas-Taussig shunt) to increase pulmonary blood flow and oxygenation.
- ❑The placement of a pulmonary artery stent can increase pulmonary blood flow.


Modified
Blalock-Taussig
Shunt
PA
AO
LA
RA
LV
RV
Ventricular
Septal Defect
Pulmonary Stenosis
(narrowing)

Prognosis

- The prognosis varies depending on the severity of the shunt and the overload of right ventricular pressure.
- Death usually occurs in young adult dogs.
- Dogs can reach the age of 5 years.
- Sudden death is common

Key points

- Congenital heart diseases are rare in the veterinary practice of companion animals.
- The clinical signs and prognosis vary depending on the severity of the anomaly (or anomalies).