

Pulmonic Stenosis

How does the heart work?

The heart is the organ responsible for pumping blood to and from all tissues of the body. The heart is divided into right and left sides. The job of the right side is to pump oxygen-deficient blood returning from the body into the lungs where fresh oxygen is collected and carbon dioxide is removed. The oxygen-rich blood returning from the lungs enters the left side of the heart where it is pumped into the aorta then to the rest of the body via the arterial system.

Each side of the heart has two chambers, an upper atrium and a lower ventricle. Between the atrium and ventricle on each side lies a valve – the tricuspid on the right and the mitral on the left – that regulates blood flow into the chambers. As the heart pumps, these valves act as one-way gates allowing blood to flow from the atrium above to the ventricle below and preventing blood from flowing back into the atrium. From the ventricles, blood is then forced to flow out into the pulmonary artery (on the right) or the aorta (on the left) through a second series of one-way valves called the pulmonic valve and the aortic valve, respectively. The region immediately before the pulmonic and aortic valves is called the ventricular outflow tract (this is the narrowest part of both ventricles).



Key:

RA: right atrium RV: right ventricle PA: pulmonic artery LA: left atrium LV: left ventricle AO: aorta



What is Pulmonic Stenosis?

Pulmonic stenosis (PS) refers to a narrowing of the pulmonic valve (stenosis is the medical term for narrowing). It is one of the most common congenital (in-born) heart diseases to affect dogs. Severe cases can create a significant problem for the affected dog, however mild cases are generally asymptomatic and live normal lives. Occasionally cats are also affected.

The most common form of pulmonic stenosis involves a deformed pulmonary valve in which the valve leaflets are too thick, the opening is too narrow, or the valve cusps are fused (or some combination of all three). The heart is forced to work harder to push the blood through the unusually narrow, stiff valve.

The consequences of pulmonic stenosis depend on the severity of the stenosis. Speaking about severe cases, the muscle of the right ventricle slowly thickens and grows due to the excess work it must perform. This thickening is initially compensatory however later it becomes detrimental to cardiac function. Eventually this thickening can cause dysfunction of the heart muscle which may lead to significant enlargement of the right side of the heart. In severe cases this can lead to right-sided congestive heart failure (meaning fluid 'backs' up in the abdomen and/or around the lungs). The other main detrimental effect of this thickening is inadequate oxygen supply to the heart muscle (blood vessels generally do not grow in proportion with the heart muscle). The result can be abnormal heart rhythms (called ventricular arrhythmias) that can cause fainting and even sudden cardiac death (albeit the latter is uncommon).

Predisposed breeds for this condition include: Terrier breeds, Miniature Schnauzers, Samoyeds, English bulldogs, Boxers, Chow Chows, Beagles, Cocker Spaniels, Basset Hounds and Mastiffs. The predisposition in these breeds suggests a genetic/heritable basis.



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What does this all mean for your pet?

Mild cases are of little concern and usually do not affect life expectancy. More severe cases however, may lead to the development of clinical signs and reduce life expectancy. Clinical signs sometimes seen include:

- Exercise intolerance, lethargy, weakness
- Blue-tinge to the gums, especially with exertion
- Fluid accumulation in the abdomen (due to right-sided heart failure)
- Fainting spells (from an abnormal electrical heart rhythm)

Severe cases that are not treated have a poor long-term prognosis (see treatment below).

How is the diagnosis made?

The turbulent blood flow resulting from the heart pumping blood through the narrowed pulmonic valve generates an abnormal heart sound called a murmur. Often, the presence of a murmur is the only clue there is a problem (clinical signs generally do not develop until later in the disease). The presence of a characteristic murmur, particularly in a young dog of a predisposed breed, increases the index of suspicion of pulmonic stenosis.

A definitive diagnosis however, can normally only be made by echocardiography (cardiac ultrasound). In addition to making a definitive diagnosis, echocardiography has many other benefits including: assessing the severity of the stenosis, the risk of congestive heart failure developing in the short-term, ruling out concurrent heart defects and providing valuable pre-operative information (if treatment is being considered).

How is pulmonic stenosis treated?

Not all dogs with pulmonic stenosis require treatment. Mild cases (and some moderate cases) generally never develop clinical problems and live a normal life. Severe cases however normally require treatment to minimise the future risk of clinical problems and increase life expectancy.

In severe cases the treatment of choice is a procedure called balloon valvuloplasty. This is a minimally invasive procedure (i.e. 'keyhole' surgery) that involves the placement of an inflatable balloon device across the narrowed pulmonic valve. This is achieved via a small incision in the neck, following which a balloon catheter is placed into the jugular vein and advanced toward the heart and the pulmonic valve under fluoroscopic guidance (real-time x-ray). Once appropriately positioned, the balloon is inflated which opens the narrowed region. The balloon may be inflated more than once before it is removed from the patient and the surgical wound closed. Because of the non-invasive nature of this surgery, post-operative recovery is short, and most dogs can go home the next day.

The success rate of this procedure depends on several variables. An important consideration is the severity of the stenosis and specific type of stenosis (which can vary considerably between individual dogs and breeds). Some dogs (e.g. Bulldogs) can get a particularly severe form of pulmonic stenosis, that may have a poorer response to balloon valvuloplasty (success may only be achieved in 50-60%)

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of such dogs). In contrast, other dogs and breeds get a form of pulmonic stenosis that is more amenable to balloon valvuloplasty (the success rate in these dogs can approach 80-90%).

Complications during the procedure are another reason that can alter the success rate. Most complications are mild and self-limiting however occasionally a major complication can occur (e.g. rupture of a major vessel) that can have catastrophic consequences. Fortunately, such major complications are rare.

Some dogs get more unusual forms of pulmonic stenosis that cannot be treated with balloon valvuloplasty. As an example, Boxers and English Bulldogs can be born with abnormalities of their coronary arteries (the vessels that supply oxygen to the heart itself), which in some cases, can 'wrap' around the pulmonic valve and create an external constriction. Balloon valvuloplasty is not performed in these patients due to the significant risk of death during the procedure.

Are there medications that can help?

There are no medications that can achieve the same outcome as balloon valvuloplasty. However sometimes medications are necessary:

- Beta blockers: These medications are designed to slow heart rate and reduce contractility. These effects are often helpful in dogs with pulmonic stenosis because they improve filling of the right ventricle as well as mildly reducing the severity of the stenosis. They are often administered for several weeks prior to balloon valvuloplasty and continued for several months afterwards.
- Diuretics: These are only necessary in advanced cases that have developed congestive heart failure. They force the kidneys to expel large volumes of water from the body (through the urine) which in turn helps to eliminate the fluid accumulating in body cavities.
- Anti-arrhythmic drugs: These are generally reserved for dogs with severe ventricular arrhythmias.

What is the prognosis?

As already mentioned the prognosis depends on the severity of the stenosis. Mild cases generally have a normal life expectancy. Moderate cases may or may not have a reduced life expectancy. Severe cases have a reduced life expectancy however with successful balloon valvuloplasty prognosis is improved (often by years).

Prognosis is best discussed with a veterinary cardiologist on a case by case basis as there are many individual variables that can alter the outcome.

Additional Resource:

http://vetmed.tufts.edu/heartsmart/

This is a very useful and well-written resource, providing pet owners with a clear and credible source of information about veterinary cardiology.

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