

Clinical Case Seminar

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An unusual case of *cortriatriumdexter* in a dog

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Abstract

Cortriatriumdexter (CTD) is a rare congenital malformation that can affect both humans and animals, in which the right atrium (RA) is divided into two chambers by a membranous structure. There is a high incidence of other cardiac congenital abnormalities associated with CTD. Clinical manifestations vary widely according to the degree of partitioning or septation of the RA. This study shows the clinical case of a two-year-old, male intact, crossbreed dog, affected from CTD associated with three other congenital heart malformations: pulmonic stenosis, mitral and tricuspid dysplasia. Diagnosis, monitoring and drug therapy of the clinical case will be reported.

Key Words: Cortriatriumdexter, cardiac defect, canine heart disease, CTD, CHD

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Introduction

Cortriatriatum is a very rare heart malformation that can affect humans (1), dogs (2) and cats (3). The *cortriatriatum* involves the division of an atrium into two smaller chambers (4,5,6). According to the atrium affected by the malformation, it is called *cortriatriatumdexter* (CTD) for the right atrium (RA), or *cortriatriatum sinister* if the left atrium is affected (4,5,6). CTD is caused by an abnormal septation of RA which is divided for the persistence of the embryonic right valve of the sinus venosus (4,5,6). This septum may range from a reticulum to a substantial sheet of tissue (4). The division of the RA causes a different pressure gradient between the small cranial sub-chamber, which receives the cranial vena cava and has extensive contacts with the tricuspid annulus, and the caudal sub-chamber which instead receives blood from the caudal vena cava (4,5,6). In most cases of CTD, the caudal vena cava is more affected by this division because the caudal sub-chamber in RA is often with a greater pressure gradient than the cranial one (4,5,6).

The abnormal membrane can cause RA obstruction of varying degrees, which depends on the blood flow and the pressure gradient between the two chambers of the RA (4,5,6). Other important factors, are the size of the membrane perforation between the two chambers and other possible associated heart defects. In fact, pulmonary stenosis, tricuspid valve dysplasia, persistent foramen

ovale, pericardial agenesis, persistent left cranial vena cava, Ebstein's anomaly and ventricular septal defect are often found in dogs with CTD (2,4).

CTD in humans has a 0.025% incidence of all congenital cardiac malformations (6), while in dogs there are no statistical estimates but only occasional case reports (2,7,8,9). In both dogs and humans, CTD can cause clinical manifestations of varying severity (1,2,6). In both species, symptoms are mainly associated with right heart failure causing: exercise intolerance, exercise-induced dyspnea, syncope, peripheral edema, pleural effusion, hepatomegaly, and cyanosis (1,2,6). Very mild or asymptomatic cases are also described in humans (1,2,6).

Clinical Case

A 2-year-old crossbreed, male, weighing 22.6 kg, with lethargy and anorexia for about 3 weeks, was referred. His vet examined the dog a week earlier and had detected a heart murmur. The dog also underwent an abdominal echo examination which showed abdominal effusion. All the parameters of the biochemical and blood count were within the physiological ranges. Then, until the cardiological examination, the dog was treated with furosemide 2 mg/kg BID, which was followed by an improvement of the clinical conditions.

One week later, at the cardiological examination the dog showed: 100 beats per minute, 30 breaths per minute, 38.5 °C temperature, pale mucous membranes and capillary refill time >2 seconds. A basilar meso-systolic heart murmurs of intensity IV/VI, with PMI on the left side was detected. The radiographic examination of chest revealed: moderate enlargement of the RA region, cardiac silhouette with VHS = 10.5 and enlargement of pulmonary veins. Electrocardiogram examination showed right axis deviation and P wave with increased amplitude. Two-dimensional, Doppler and color Doppler echocardiography was performed with an ultrasonographic machine (M9Vet, Mindray, China), and revealed multiple congenital heart defects. A right atrial enlargement and a dilated coronary sinus has been found. The echocardiographic examination showed a perforated membrane divided the RA into two chambers suggestive of CTD (Fig. 1), this finding was also confirmed by agitated saline contrast. The tricuspid valve was dysplastic for the presence of thickened and shortened leaflets, with limited motion and inadequate coaptation during the systole (Fig. 1). The mitral valve was also abnormal, had thickened leaflets and showed prolapse during ventricular systole (Fig. 1). The left atrium appears enlarged, in fact it showed a left atrium-aorta ratio score of 1.9 (reference ranges <1.6) (Fig. 2) (10). The pulmonary valve annulus was hypoplastic with an infundibular restricting and thickened motionless leaflets (Fig. 3). The blood flow through the pulmonary valve was turbulent, with a peak velocity of 3.7 m / s, suggesting a peak systolic transvalvular pressure gradient of 53 mmHg. The dog was therefore affected by pulmonary stenosis (11) (Fig. 3). The final diagnosis was CTD, associated with pulmonic stenosis,

mitral and tricuspid dysplasia. An angiography was proposed to better characterize the membrane inside the RA and therefore plan a surgery (8,11) but the owner refused any other non-medical treatment. Then, the dog continued furosemide therapy at 2 mg/kg/os/BID and showed an excellent initial response. In fact, at the follow-up visit performed 2 weeks after the start of treatment with furosemide, the abdominal effusion had decreased and the dog had a further improvement in clinical conditions. Despite the therapy, however, the dog showed clinical worsening caused by large pleural and abdominal effusion two months later. Therefore, the dog was euthanized on the owner request. The autopsy was proposed to the owner but he refused.

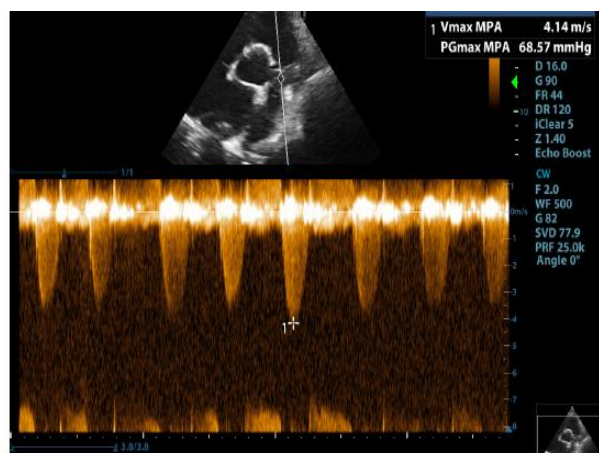
Fig. 1. A perforated membrane dividing the right atrium suggestive of CTD is present. The tricuspid valve and the mitral valve are displasic.



Fig. 2. The left atrium appears enlarged. The ratio between the left atrium and the annulus aortic is 1.9



Fig. 3. The pulmonary valve annulus is hypoplastic with a turbulent blood flow suggestive of moderate pulmonary stenosis



Discussion and conclusion

CTD is a congenital disease that causes a heart with three atria (1,5). The division of RA can have different degrees resulting in clinical manifestations of different severity (2,4). CTD is also often associated with other congenital heart defects (4,11). The embryologic mechanism behind CTD, and the relationship between CTD and other congenital heart defects, remains not fully understood (4,11). It is possible that insufficient apoptosis during the formation of atrioventricular or semilunar valves

could cause an incomplete separation of the tricuspid valve leaflet from the right ventricular wall, and the persistence of sinus venosus valve (12,13). Genetic mutations occurring during pregnancy may also have an important role in the development of this defect (12,13), but further studies are necessary to better understand the origin of the CTD.

In this case, a dog with CTD with associated moderate pulmonic stenosis, tricuspid and mitral dysplasia is described. Anorexia and lethargy are non-specific clinical findings, but ascites and cardiac murmur suggest right-sided heart disease. Although the radiographic examination is not specific for CTD, the enlargement of the cardiac silhouette, the increase size of the pulmonary veins and the enlargement of the RA are radiographic findings associated with right-sided congestive heart failure and require an evaluation of the heart by echocardiographic examination (11). In fact, the echocardiographic examination is considered the most useful tool for the diagnosis, evaluation and monitoring of CTD and other heart diseases (4,11). However, it should be emphasized that it is necessary to use the saline-agitated contrast to differentiate CTD from aneurysmal dilation of the RA (2,4). In the literature there is debate on the prognostic utility of the peak diastolic velocity of the transmembrane flow between the two small chambers within the RA (2). In dogs with CTD without other heart defects, the peak diastolic velocity of the transmembrane flow is likely to reflect the severity of the obstruction. In this case report, the CTD was associated with other right-sided cardiac malformations, such as tricuspid dysplasia and pulmonary stenosis, which influenced the pressure gradient between the two chambers and therefore the measurement was unreliable (2).

The best treatment for dogs with CTD is surgery (2,4,8,9). Interventional procedures including the balloon membranostomy, stent implantation or surgical resection, when performed, present a good success in improving clinical signs and no serious complications are reported, but they are to prefer in dogs that show mild clinical signs in absence of pulmonic hypertension (2,4,8,9). When surgical treatment is not possible, drug therapy improves the dog's quality of life but the long-term prognosis is not good (4,11).

Diuretic treatment decreases pressure in the RA and the formation of abdominal effusion. Other drugs to be added to furosemide must be evaluated according to the haemodynamic effects that the other heart defects associated with CTD have and therefore the echocardiographic examination have a very important role (2,4,11). In our case, for example, if the pulmonary stenosis had been severe instead of moderate, the addition of beta-blockers could have been evaluated to decrease the dynamic obstruction of the outflow tract and the hypertrophy of the right ventricle (11). An important class of drugs in the long-term therapeutic management of atrioventricular valvular insufficiency are angiotensin-converting enzyme inhibitors (ACEI) (11,14). In fact, ACEIs decrease the effects of the renin-angiotensin-aldosterone system (RAAS), lower blood pressure and increase long-term survival (11,14).

The role of ACEIs in acute treatment is less clear (14).

In conclusion, it is important to keep in mind that each medical therapy must be adapted to the specific clinical case and its unique characteristics. It should also be noted that all dogs treated with drug therapy must be carefully monitored periodically to avoid electrolyte, blood pressure, haemodynamic imbalances and increased renal values (11,14).

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References

1. Jha, A.K., Makhija, N. (2017). CorTriatriatum: A Review. *SeminCardiothoracVascAnesth*, 21(2), 178-185. doi: 10.1177/1089253216680495.
2. Nadolny, K.E., Kellihan, H.B., Scansen, B.A., Tjostheim, S.S., Grint, K.A., Forrest, L.J., Stepien, R.L. (2019). Cortriatriatumdexter in 17 dogs. *J Vet Cardiol*, 23, 129-141. Doi: 10.1016/j.jvc.2019.03.002.
3. Koie, H., Sato, T., Nakagawa, H., Sakai, T. (2000). Cortriatriatum sinister in a cat. *JSAP*, 41, 128-131. <https://doi.org/10.1111/j.1748-5827.2000.tb03180.x>
4. Ettinger, S.J., Feldman, E.C. (2005). *Textbook of Veterinary Internal Medicine*. 6th Ed. Elsevier Saunders, St. Louis, USA. Pp.1288-1289.
5. Bharucha, T., Spicer, D.E., Mohun, T.J., Black, D. (2015). Cortriatriatum or divided atriums: which approach provides the better understanding? *Cardiol Young*, 25, 193-207. doi: 10.1017/S1047951114000572.
6. Moral, S., Ballesteros, E., Huguete, M., Panaro, A., Palet, J., Evangelista, A. (2016). Differential diagnosis and clinical implications of remnants of the right valve of the sinus venosus. *J Am SocEchocardiogr*. 29(3), 183-94. Doi: 10.1016/j.echo.2015.11.018.
7. Yang, V.K., Nussbaum, L., Rush, J.E., Cunningham, S.M., MacGregor, J., Antoon, K.N. (2015). Congenital cardiac malformation with three-chambered right atrium and a persistent left cranial vena cava in a dog. *J Vet Cardiol*, 17, 62-70. Doi: 10.1016/j.jvc.2014.12.002.
8. Tanaka, R., Hoshi, K., Shimizu, M., Hirao, H., Akiyama, M., Kobayashi, M., Machida, N., Maruo, K., Yamane, Y. (2003). Surgical correction of cortriatriatumdexter in a dog under extracorporeal circulation. *J Small AnimPract*. 44(8), 370-3. doi: 10.1111/j.1748-5827.2003.tb00171.x.
9. Chanoit, G., Bublot, I., Viguier, E. (2009). Transient tricuspid valveregurgitation following surgical treatment of cortria-triatumdexter in a dog. *J Small AnimPract*, 50, 241-5. doi: 10.1111/j.1748-5827.2008.00700.x.
10. Rishniw, M., Caivano, D., Dickson, D., Vatne, L., Harris, J., J.N. Matos, J.N. (2019). Two-dimensional echocardiographic left-atrial-to-aortic ratio in healthy adult dogs: a reexamination of reference intervals. *J Vet Card*, 26, 29-38. doi: 10.1016/j.jvc.2019.11.001.
11. Santilli, R., Bussadori, C., Borgarelli, M. (2018). *Manual of Cardiology: Dog and Cat*. 3rd Ed. Edra, Milano, Italia.
12. James, T.N. (1998). Normal and abnormal consequences of apoptosis in the human heart. *Annu Rev Physiol* 60, 309-325. Doi: 10.1146/annurev.physiol.60.1.309.
13. Lamers, W.H., Moorman, A.F.M. (2002). Cardiac septation: a late contribution of the embryonic primary myocardium to heart morphogenesis. *Circ Res*, 91, 93-103. Doi: 10.1161/01.res.0000027135.63141.89.
14. Keene, B.W., Atkins, C.E., Bonagura, J.D., Fox, P.R., Häggström, J., Fuentes, V.L., Oyama, M.A, Rush, J.H., Stepien, R., Uechi, E. (2019). ACVIM consensus guidelines for the diagnosis and treatment of myxomatous mitral valve disease in dogs. *J Vet Intern Med*, 33, 1127-1140. doi: <https://doi.org/10.1111/jvim.15488>



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