Cor triatriatum dexter of unusual morphology in a miniature schnauzer

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Abstract

A five-year-nine-month-old, male entire, miniature schnauzer presented for further investigation of pleural effusion. Echocardiography revealed a perforated membrane dividing the right atrium into two chambers: the true right atrium (a small, lower pressure, cranioventral chamber communicating with the tricuspid valve and right ventricle) and the accessory right atrium (a larger, higher-pressure, caudodorsal chamber), consistent with a cor triatriatum dexter (CTD). This was confirmed on computed tomography angiography. Imaging studies revealed that both the cranial and caudal vena cava entered the higher-pressure accessory right atrium and the coronary sinus entered both the accessory and true right atrial chambers. This differed from the more usual canine CTD presentation with the cranial vena cava entering the lower-pressure cranial chamber and the caudal vena cava entering the higher-pressure caudal chamber. Balloon membranostomy was successful in reducing the pressure gradient between the two right atrial chambers with subsequent resolution of the clinical signs. The patient continues to do well after three-years of follow-up.

Keywords

Congenital heart disease, congestive heart failure, echocardiography, canine

Abbreviations

CdVC	caudal vena cava
CrVC	cranial vena cava
CS	coronary sinus
CTA	computed tomography angiography
CTD	cor triatriatum dexter
RI	reference interval
RA	right atrium
VTI	velocity time integral

A five-year-nine-month-old, male entire, miniature schnauzer was referred to the Small Animal Teaching Hospital for further investigation of pleural effusion. The patient had been treated at the referring practice for suspected pancreatitis, following intermittent abdominal bloating, diarrhoea, and tachypnoea of one-year duration. Referral was sought after three weeks of progressive lethargy and tachypnoea; the patient had an in-dwelling thoracic drain placed three days prior to referral (modified transudate drained) and received furosemide.

On presentation the dog was lethargic, weighing 7.7 kg, body condition score 4/9. Mucous membranes were pink with a normal capillary refill time. Heart rate was 80 beats/minute with a regular rhythm and good quality, synchronous pulses. Respiratory rate was 50 breaths/minute with mildly increased inspiratory effort. No adventitious lung sounds or murmur, were auscultated. Abdominal palpation was hindered due to the patient's nervous demeanour. There was jugular distension and

pulsation and a positive hepatojugular reflux (indicative of elevated pressure within the right atrium). The rest of physical examination was unremarkable.

Doppler systolic blood pressure was normal (135 mmHg, reference interval [RI] 110-160 [1]). Haematology was unremarkable. Serum biochemistry showed mildly elevated urea (8.2 mmol/L, RI 3.5-6.0), creatinine (124 µmol/L, RI 20-110; likely pre-renal secondary to furosemide) and ALT (64 U/L, RI 7-50; possible hepatic congestion).

Thoracic ultrasound revealed moderate volume pleural effusion; 230 mLs was drained using the in-dwelling thoracic drain. Echocardiography[a] revealed a thick hyperechoic membrane dividing the right atrium (RA) into a large, accessory RA and smaller, more cranial, true RA which communicated with the right ventricle (Supplement Fig. I). Spontaneous echo-contrast was seen within both RA chambers, suggestive of blood stasis. Colour flow Doppler showed a continuous jet of blood flow from the accessory RA to the true RA with a peak end-diastolic velocity of 2 m/s and a velocity time integral (VTI) of 101.9 cm. The coronary sinus (CS) was subjectively dilated, appearing to join the accessory RA. The interatrial septum bowed to the left, with no blood flow demonstrated across this on colour-flow Doppler. The tricuspid valve apparatus and tricuspid inflow appeared normal with no tricuspid regurgitation. The left heart was unremarkable. To better understand the communication between the RA chambers, three echocontrast (bubble) studies using agitated saline mixed with colloid[b] were performed via the left saphenous, left cephalic and right cephalic vein (Video 1-3). In all studies, bubbles entered the accessory RA, before entering the true RA; no atrial septal defect or other shunting of blood was seen and no contrast entered via the CS, excluding the presence of a persistent left cranial vena cava (CrVC). These findings were considered consistent

with a perforated cor triatriatum dexter (CTD) with both caudal vena cava (CdVC) and CrVC flow entering the larger, high pressure, accessory RA.

Considering the patient's older age at presentation, lack of ascites and concurrent evidence of cranial right-sided congestive heart failure, which did not meet criteria for typical CTD [2], further investigations were pursued to better define the defect. Computed tomography angiography (CTA) was performed with an 80-slice CT scanner[c] using retrospective ECG-gating. A bolus of iodinated contrast medium[d] was injected via pressure injector[e]. The CTA showed both the CdVC and CrVC to be severely enlarged, joining the larger, caudodorsal, high pressure, RA chamber (Fig. 1 and 2; Video 4). The smaller, cranioventral chamber was continuous with the right atrial appendage. The CS was subjectively dilated and entered both RA chambers (Fig. 2); the azygous vein was also mildly distended. The membrane dividing the RA was perforated, or very thin, in several places (Fig. 1 and 2). A diverticulum-like structure was seen at the region of the ductus arteriosus; a membrane was visible between this and the main pulmonary artery with no communication between the two. Abdominal CT showed hepatomegaly and marked hepatic venous distension.

The CTA supported the echocardiographic findings, the final diagnosis being a perforated CTD. Medical management with furosemide (1 mg/kg q8h IV), benazepril/spironolactone (2.5/20 mg PO q24h) and clopidogrel (18.75 mg PO q24h) was initiated; the latter due to the spontaneous echo-contrast and perceived risk of thromboembolic complications. Two days post-admission, pleural effusion had not recurred, and the thoracic drain was removed. The patient was discharged the following day on furosemide (2mg/kg PO q12h) and the above oral medications.

Two weeks later the dog presented for membranostomy of the CTD via balloon dilation. Lung sounds were reduced ventrally bilaterally, thoracic ultrasound confirming pleural effusion. Under general anaesthesia[f] a Mila thoracic drain[g] was placed (450 mLs pleural fluid drained). The dog was maintained in right lateral recumbency. Fluoroscopic guidance and intra-operative transoesophageal echocardiography were utilised. Both the left jugular vein and right femoral vein were visualised via a cut-down approach, and then catheterised[h], through which catheters[ijk] were advanced into the CrVC and CdVC respectively. Pressure in the CrVC measured 15 mmHg. Angiography via the CrVC (Video 5) and CdVC (Video 6) was performed using 1 mL/kg iodinated, non-ionic contrast medium[lm] each time. These confirmed entry of both vena cavae into the high-pressure accessory RA and aided in localisation of the membrane ostium. Advancement of a multipurpose catheter[j] through the ostium of the RA membrane was only possible via the CrVC approach (too acute an angle with the ostium and the catheter via the CdVC). The catheter was replaced by a 16 mm x 4 cm balloon catheter[n], (the largest size possible considering the diameter of the vascular introducer), which was inflated three times at the level of the ostium; complete loss of the balloon waist was achieved on the third inflation. The velocities through the ostium were measured by transoesophageal spectral Doppler intra-operatively, reducing from 1.15 m/s to 0.5 m/s; the 2D and colour flow images also showed a wider communication between the two RA chambers.

Echocardiography the following day showed a smaller accessory RA and collapse of the membrane, suggesting reduced pressure. The true RA subjectively appeared better filled. The velocity across the CTD membrane reduced to 1.1 m/s, consistent with a reduced pressure gradient between the two right atrial chambers. The VTI

reduced to 42.5 cm; when the VTIs were divided by the R-R intervals (to account for the difference in heart rates pre and post ballooning), the pre-VTI was 0.1 cm/ms, and the post-VTI was 0.061 cm/ms, equating to a 39% decrease in VTI after ballooning. Repeat bubble study subjectively showed improved flow across the CTD membrane, with bubbles flowing more freely and quickly from the accessory to the true RA. No recurrence of pleural effusion was seen. The dog was discharged two days post-operatively on furosemide (1 mg/kg PO q12h), and benazepril/ spironolactone. The dog was stable at two-week recheck with no pleural effusion and no hepatojugular reflux. Oral medications were discontinued over the following six-months. Echocardiography at six-months showed similar right-sided changes as post-operatively, with reduced pressures within the accessory RA and no effusion. The dog continues to do well requiring no medications three years post-operatively.

Discussion

Although CTD is well documented in dogs [3], this case represents an anatomical variation, which as far as the authors are aware, has not previously been reported in dogs.

Cor triatriatum dexter is a congenital defect characterised by division of the RA into cranial and caudal chambers. During embryogenesis the larger right valve of the sinus venosus becomes prominent and divides the RA into two chambers, diverting oxygenated venous blood from the CdVC to the left heart via the foramen ovale [4]. By around 12 weeks of gestation (in humans) this valve will normally have significantly regressed, the remnants becoming the crista terminalis cranially and the Eustachian valve (adjacent to the caudal vena cava) and Thebesian valve (adjacent

147 to the orifice of the coronary sinus) caudally. Failure of this regression leads to
148 persistent RA compartmentalisation [3,5,6].

149 Four morphological variations of CTD in people have been proposed: true CTD (with
150 demonstration of obstructed blood flow), Chiari-like malformation (a reticular network
151 of fibres, common in people), residual Eustachian valve and residual Thebesian
152 valve, the latter two not causing significant obstruction to blood flow [6]. A true CTD
153 can be further divided into four types based on position of the membrane in relation
154 to the venous inflow to the right heart (Fig. 3) [7,8]. Previously reported canine cases
155 [9,10,11,12,13,14] are most analogous with a type III CTD, with the CrVC entering
156 the true, cranial chamber and the CdVC (plus/minus the coronary sinus) entering the
157 caudal accessory chamber, thus resulting in caudal signs of right-sided heart failure
158 (predominantly ascites); many of these cases also reported concurrent, right-to-left
159 shunting, patent foramen ovale. Dobak *et al.* [15] hypothesised that failure of
160 regression of only the more caudal portion of the right sinus venous valve, causes
161 this type III morphology with obstruction to only the CdVC flow. However, in this
162 case, both the CrVC and CdVC enter the caudodorsal accessory chamber, most
163 analogous with a type II CTD (Fig. 3), which explains the presence of cranial signs of
164 right-sided failure. Dennler *et al.* [16] reported a case of CTD where the CrVC
165 appeared obstructed due to an aberrant azygous vein, the flow immediately entering
166 both cranial and caudal RA chambers, however, no pressure gradient could be
167 proven on invasive measurements.

168 Medical management of CTD is frequently unsuccessful [17]; surgical intervention
169 thus being the mainstay of treatment. Balloon dilation is well documented for the
170 treatment of canine CTD [11,12]; stent placement and use of cutting balloons has
171 also been described [17,18]. In this case balloon dilation was successful in reducing

172 the pressure gradient between the two RA chambers, with a subsequent resolution
173 of right-sided congestive heart failure and resolution of clinical signs. A cutting
174 balloon would have been considered should an intra-operative improvement not
175 have been seen.

176

177 It is more common for cases of canine CTD to present at a young age, although
178 there are reports of the diagnosis being made in mature dogs [11,14,16], as in this
179 case. The haemodynamic significance of CTD is dependent on the severity of the
180 obstruction (determined by whether the membrane is perforate or imperforate and
181 the size of the membrane orifice in the former). Thus, in cases of perforate CTD with
182 a large enough orifice and minimal obstruction to blood flow, there may be only mild
183 increases in right atrial pressures and as such clinical signs may not develop; this
184 likely explaining the delayed onset of clinical signs in this case. The absence of a
185 concurrent patent foramen ovale (and secondary hypoxaemia) may also have
186 delayed the development of clinical signs. An explanation as to why this dog
187 converted from a previously asymptomatic to a symptomatic state is speculative. In
188 adult humans with cor triatriatum sinister (compartmentalisation of the left atrium)
189 fibrosis of the dividing membrane (with or without calcification) has been proposed
190 as a mechanism for late onset clinical signs [19]; this could be an explanation in this
191 case, fibrosis reducing the orifice area with resultant signs of right-sided failure.

192 In conclusion, variations in the anatomy and subsequent clinical signs are possible in
193 dogs with CTD; this case representing a rare morphological variation with both the
194 CrVC and CdVC entering the accessory RA chamber, thus explaining the presence
195 of both cranial and caudal signs of right-sided congestive heart failure.

Conflicts of interest statement

The authors have no conflicts of interest

Footnotes

[a] GE Vivid 7, GEHeathcare, Buckinghamshire, UK.

[b] Gelofusine 4%, Braun, Australia.

[c] Aquilion Prime, Toshiba Medical Systems Europe, The Netherlands.

[d] 600mg I/kg, Xenetix, iobitriol, 300mg/mL, Guerbet, France.

[e] Medrad®Stellant® CT Injection System, Bayer, Germany.

[f] Pre-medication: buprenorphine (20 µg/kg IV) and medetomidine (2 µg/kg IV). Induction: midazolam (0.2 mg/kg) and propofol to effect. Maintenance: sevoflurane in oxygen and medetomidine constant rate infusion. Intra-operative cefuroxime (15mg/kg IV q90minutes intra-operatively).

[h] MILA International Inc. Kentucky, USA.

[h] 7 and 6 French Radiofocus introducer, Terumo, 3001 Leuven, Belgium.

[i] Stiff and standard 0.025" x 260 cm Radiofocus guidewire, Terumo, 3001 Leuven, Belgium.

[j] 4 French Multi-purpose angiographic catheter, AngioDynamics, Latham, New York, USA.

[k] 4 French Occlu-Marker Pigtail Catheter, PFM medical, Poynton, Cheshire, UK.

[l] Optiray 350 mg/mL at 1.5 mLs/s, Ioversol, Mallinckrodt/Tyco healthcare, Hazelwood, MO, USA.

[m] Avida® angiographic contrast injector, Imaxeon, Australia.

[n] Tyshak II Veterinary Balloon catheter, Infiniti Medical, Redwood City, CA, USA.

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Figures

Fig. 1. Parasagittally reconstructed CT image of the thorax post contrast administration through a plane including the cranial and caudal vena cava, right atrium, and ventricle. A membrane is visible dividing the right atrium into a larger caudodorsal (white asterisk) and smaller cranioventral chamber (white block arrow) which is extending medially as the right auricle (not visible in this image). The membrane is perforate/very thin at the cranial aspect (small white arrow). The coronary venous sinus is noted entering the caudodorsal chamber (open arrow). Cranial and caudal vena cava are distended.

Fig. 2. Computed tomography image of an oblique reconstruction of the thorax post contrast administration parallel to the heart base. The cranioventral chamber (✶) is visible cranially, including part of the right auricle (+). The caudodorsal, accessory, chamber (black open arrow) is divided from the cranioventral, true, chamber by a perforate curvilinear membrane. The coronary venous sinus is seen to enter the caudodorsal chamber and cranioventral chamber (black block arrow).

Fig. 3. Schematic diagram illustrating the varying morphologies of CTD determined by the position of the membrane with respect to the venous inflow to the right heart as reported in 316 people. For comparison to the present case, the example is shown without a patent foramen ovale and a perforate membrane. The membrane divides the right atrium into two chambers, a caudodorsal accessory chamber (*) and a cranioventral true chamber (+) which communicates with the right ventricle. In type

I CTD the membrane is positioned so that the cranial vena cava (black block arrow), caudal vena cava (open block arrow) and coronary sinus (filled in circle) all enter the caudodorsal chamber. In type II CTD both the cranial and caudal vena cava enter the caudodorsal chamber, while the coronary sinus enters the cranioventral chamber. In type III CTD only the caudal vena cava enters the caudodorsal chamber. In type IV CTD only the coronary sinus enters the caudodorsal chamber. Image adapted from Doucette and Knoblich [7] and Hausdorf et al [8].

Supplementary material:

Figure	Title	Description
I	Echocardiography at presentation, right parasternal long axis views	There is a thick, hyperechoic membrane across the right atrium (arrow), forming two right atrial chambers, the interatrial septum bows to the left, indicating elevated right atrial pressures. There is also a residual pleural effusion. AccRA: accessory right atrium; LA: left atrium; LV: left ventricle.
Video		
1	2D echocardiography in right parasternal long axis view with bubble study via the left saphenous vein.	Bubbles illustrate flow entering the larger accessory right atrium, before entering the smaller true right atrium.

		AccRA: accessory right atrium; TrueRA: true right atrium.
2	2D echocardiography in a right parasternal oblique long axis view optimised for the right atrium with a bubble study via the left cephalic vein.	Bubbles illustrate flow from the cranial vena cava entering the larger accessory right atrium, before entering the smaller true right atrium. AccRA: accessory right atrium; TrueRA: true right atrium.
3	2D echocardiography in a right parasternal long axis view with a bubble study via the right cephalic vein.	Bubbles illustrate flow entering the larger accessory right atrium, before entering the smaller true right atrium. AccRA: accessory right atrium; TrueRA: true right atrium.
4	Sagittal thoracic angiographic computed tomography.	Both the caudal and cranial vena cava are severely enlarged, both joining in the larger, high pressure, accessory right atrial chamber
5	Fluoroscopic angiography with manual administration of contrast medium into the cranial vena cava	A pigtail catheter is positioned in the cranial vena cava. The contrast medium enters the higher-pressure accessory right atrial chamber with mild retrograde flow into the caudal vena cava. The transoesophageal ultrasound probe is seen, as well as

		a table artefact (effacing the diaphragm).
6	Fluoroscopic angiography with manual administration of contrast medium into the caudal vena cava	A pigtail catheter is positioned in the caudal vena cava. The contrast medium enters the higher-pressure accessory right atrial chamber with moderate retrograde flow into the cranial vena cava. The transoesophageal ultrasound probe is seen, as well as a table artefact (effacing the diaphragm).

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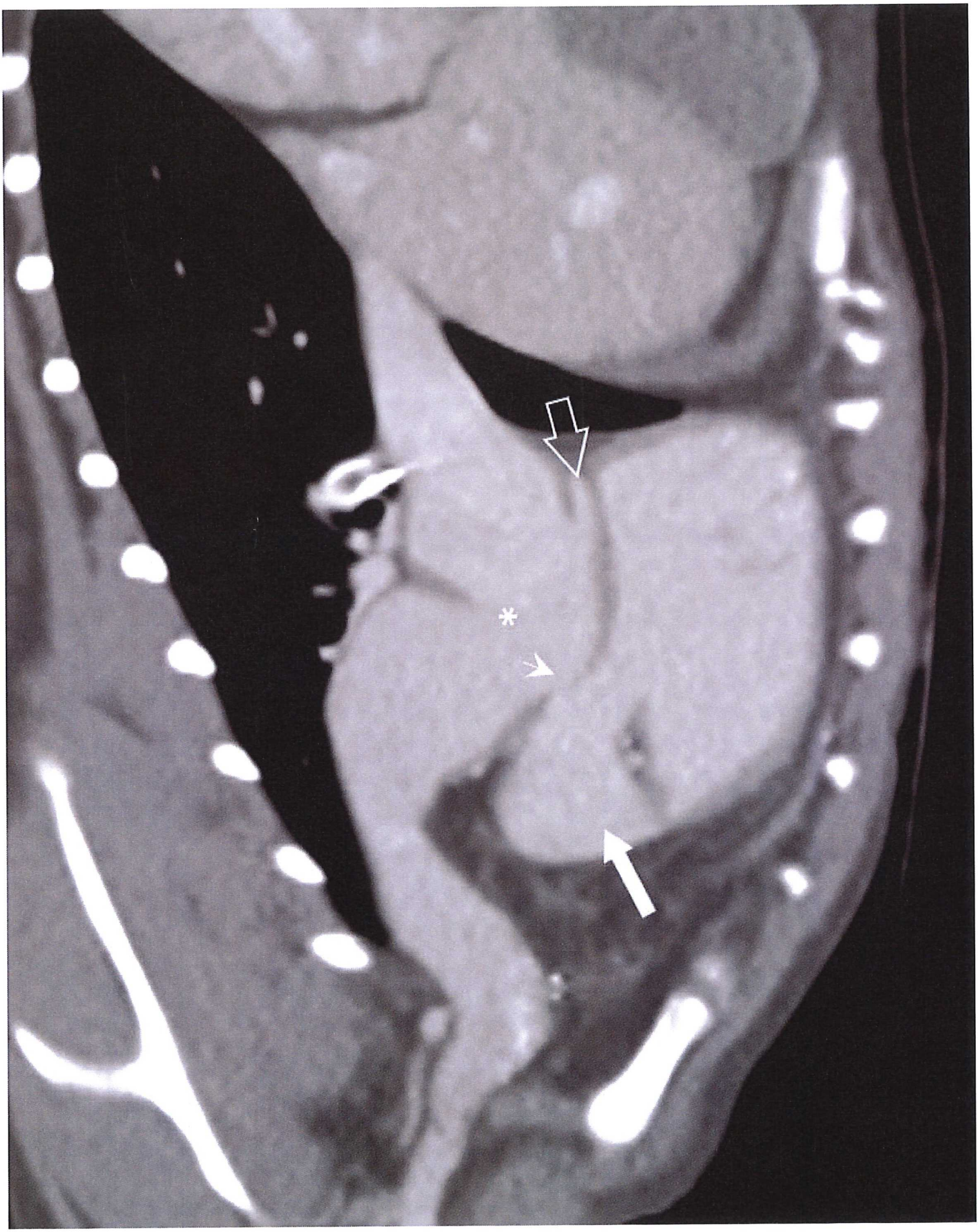

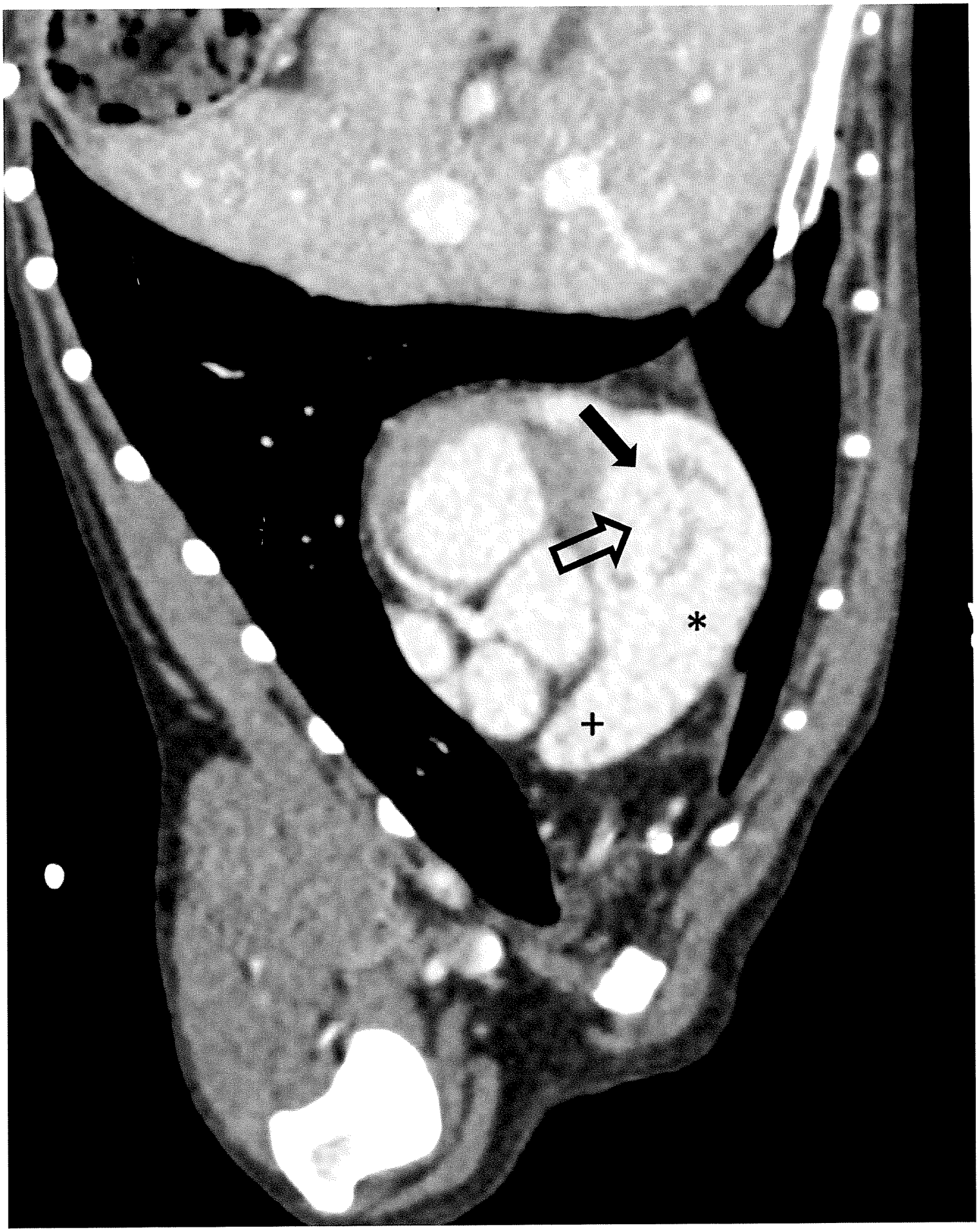

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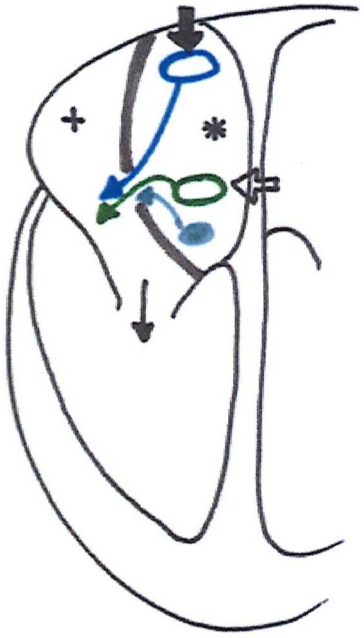

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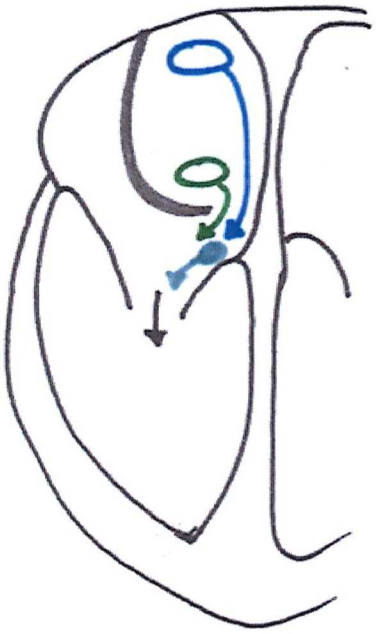

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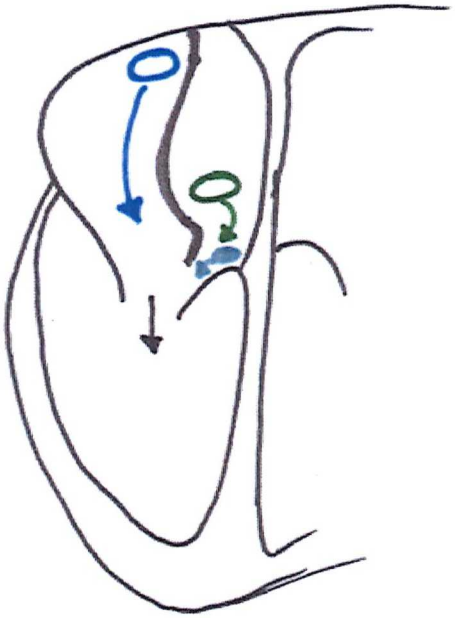

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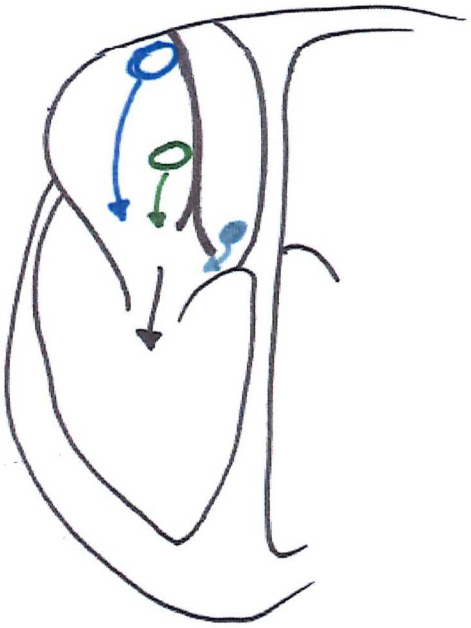

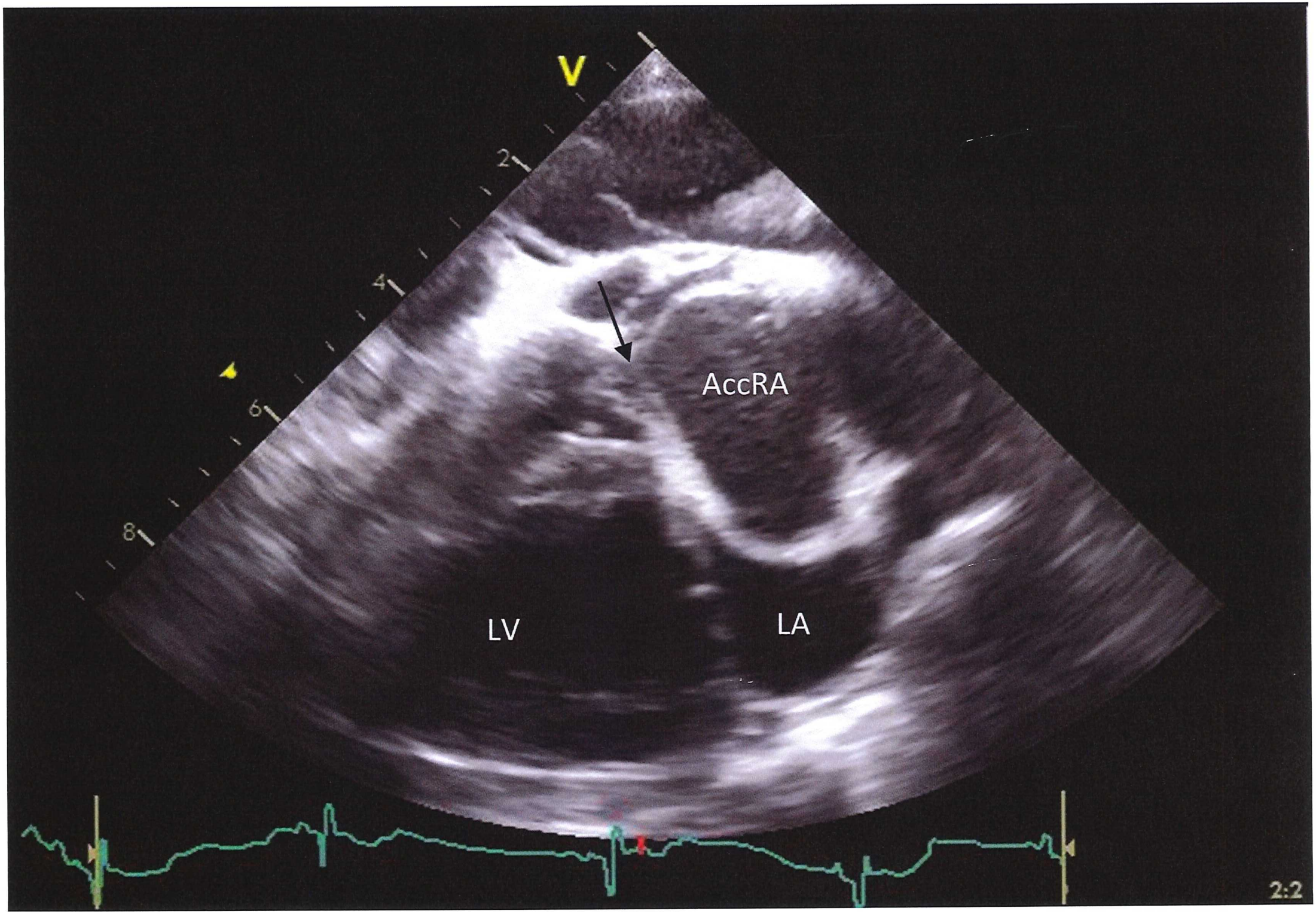
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