A case of a complete atrioventricular canal defect in a ferret

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Abstract

Background: Atrioventricular canal defect is a rare congenital disorder of the heart and describes the presence of an atrial septal defect, a variable presentation of ventricular septal alterations including ventricular septal defect malformations in the mitral and tricuspid valves. The defect has been described in human beings, dogs, cats, pigs, and horses.

Case presentation: This paper describes the case of a complete atrioventricular canal defect in a four-year-old intact male pet ferret (Mustela putorius furo), which was presented due to posterior weakness, ataxia, and decreased appetite. A loud systolic murmur, dyspnea, and hind limb paraparesis were detected during the clinical examination. Thoracic radiographs showed generalized cardiomegaly and lung edema. ECG showed sinus rhythm with prolonged P waves and QRS complexes. Echocardiography showed a large atrial septal defect, atrioventricular dysplasia, and a ventricular septal defect. Palliative treatment with oxygen, furosemide, spironolactone, enalapril, diltiazem, and supportive care was chosen as the therapy of choice. The ferret recovered gradually during hospitalization. A follow-up examination at three and six months showed stabilization of cardiac function.

Conclusions: To the authors knowledge, this is the first time an atrioventricular canal defect has been described in a pet ferret.

Keywords: Atrioventricular canal, Pet ferret, Endocardial cushion defect, Heart failure, Echocardiography

Background

An atrioventricular canal defect (AVCD) is a rare congenital disorder of the heart characterized by an ostium primum atrial septal defect (ASD), alterations of the ventricular septum, like ventricular septal defect (VSD), and defects in the atrioventricular valves (AV) [1]. It occurs due to the failure of the development of the endocardial cushions during embryogenesis and the persistence of a primitive single AV canal [2].

An AVCD may be partial (incomplete), transient or intermediate, transitional or complete. The difference between them also reflects the severity of the defect [1–4]. The complete variety results from concurrent large ASD and VSD (i.e. common canal) as well as AV dysplasia. Typically, there is a large (uninterrupted) communication between all four heart chambers. A transient AVCD has common AV that are often inserted into the IVS that partially close the gap between the chambers. The VSD is often small with a moderate restriction of blood flow. An incomplete AVCD occurs when the flaps of the AV valves are connected to an interatrial or IVS without an identifiable VSD [1, 3].

In veterinary medicine, AVCD has been described in several species: eight pigs [5], 31 cats [1, 6–9], nine dogs [2–4, 10–14], and four horses [15–18]. Factors like oxygen deficiency, viral infection during early pregnancy, nutritional, genetic, and teratogenic factors have been suggested as possible factors triggering the development of this condition and possibly other congenital heart
diseases [14, 18]. To the author’s knowledge, this is the first case of an incomplete AV canal in a ferret that has survived six months after the diagnosis with conservative therapy.

Case presentation
A four-year-old intact male pet ferret (*Mustela putorius furo*) was presented to the Avian and Exotic Animal Clinic at the University of Veterinary and Pharmaceutical Sciences Brno, Czech Republic with a seven-day history of posterior weakness, ataxia, and decreased appetite. Clinical examination revealed lethargy, hind limb paraparesis, prolonged capillary refill time (three seconds), labored respiration, and dental tartar. A marked cardiac systolic murmur (grade 5/6) was found during thoracic auscultation. Jugular vein distention was not evident. Occasional splitting of the 1st heart sound was also noted, and abdominal palpation revealed splenomegaly. Complete cell blood count, plasma biochemistry, and cardiological examination that included thoracic radiography, electrocardiography (ECG), and echocardiography were performed. Laboratory analysis showed mild hyperphosphatemia and slightly increased blood urea concentrations. Radiographs showed an enlarged cardiac silhouette (vertebral heart score 8; reference range 3.75 to 4.07) [19], bronchial pulmonary pattern, and venous congestion (Fig. 1a, b). An ECG taken in right lateral position recorded sinus rhythm with possible left atrial and left ventricular enlargement (Fig. 1c, d).

Echocardiographic examination was performed from all standard views, using an Aloka Prosound alpha 7 ultrasound machine (Hitachi Aloka Medical, Tokyo, Japan) equipped with a phased array 2–9 MHz cardio probe. Hypertrophy of the left ventricular posterior wall (diastole 6.1 mm), hypertrophy of papillary muscles and eccentric right ventricular hypertrophy with marked myocardial hypertrophy of the right ventricular wall (Fig. 2) were revealed from a right parasternal short axis view. Also, abnormal tricuspid leaflets were observed.

Right parasternal long axis and modified left apical views revealed a single very large atrium due to a large ostium primum ASD, a left-to-right VSD, a common AV that inserts in the crest of the IVS, and abnormal morphology of the tricuspid valve that displayed one leaflet abnormally prominent arising from the free wall of the right ventricle (Fig. 3a). The mitral valve seemed normal. Color flow mapping (CFM) demonstrated tricuspid and mitral insufficiency, and blood flowing
between the left and right part of the single atrium (Fig. 3b, c). The Qp / Qs determined by measuring Doppler-derived aortic and pulmonary integral systolic flow rates was 1.6, indicating increased right cardiac output. Continuous-wave Doppler analysis of the tricuspid jet revealed severe pulmonary arterial hypertension with a peak velocity of 4.39 m/s (Fig. 3d). A diagnosis of a complete AVCD Rastelli type A defect was determined based on the echocardiographic findings, which confirmed the presence of a large ostium primum ASD, VSD and dysplastic AV valves.

Initial therapy consisted of oxygen therapy and intravenous isotonic fluids. After final diagnosis, medical therapy was started with furosemide (4 mg/kg IV, QID), spironolactone (12.5 mg/kg PO, BID), enalapril (0.5 mg/kg PO, SID) and diltiazem (7.5 mg/kg PO, BID). Two days after, the patient was discharged from hospitalization with the above-mentioned therapy, except the furosemide that was given orally (4 mg/kg q24h). At a two-week follow-up, the ferret’s condition had improved and was able to walk and eat normally. Administration of furosemide and diltiazem was discontinued after two weeks and five weeks,
Discussion and conclusions

Complete AVCD is a very rare finding in all mammal species but, according to literature, may be found relatively common in cats and pigs. One paper reports about 8 cases in porcine that can be assumed as a complete AVCD or at least having the intermediate type, where interatrial and interventricular defects were frequently accompanied by other defects like subaortic or pulmonic stenosis, vegetative endocarditis, or persistent left anterior vena cava [5]. Some cats were reported to have complete AVCD Rastelli type A and B [1]. Other 26 feline patients were diagnosed as incomplete AVCD in some instances (8/26) associated with other congenital heart abnormalities [1, 6–9]. Additionally, abnormalities of the tricuspid valve similar as to what observed in the presented case were observed in cats, where a large leaflet of the valve was more prominent [7]. Dogs were reported mainly with incomplete AVCD and, strikingly, in much lesser proportion than cats [2–4, 10–14]. In horses, only complete AVCD cases were described [15–18]. To date, there is no published information about the incidence of this defect in exotic companion mammals.

The development of clinical signs depends on the morphology of the heart defect, its size, and pressure gradient between the left and right side. Generally, large defects result in marked right ventricular eccentric hypertrophy because the right ventricle is more compliant than the left ventricle in accommodating more blood volume if there is no interatrial septum [9]. This often may cause shunt reversal and poor prognosis. In this patient, there were several clinical signs of heart failure mainly due to the complete characteristics of AVCD. In general, incomplete AVCD does not cause clinical signs unless marked mitral valve regurgitation develops [1]. If heart failure is present, arrhythmias, respiratory difficulty due to pulmonary edema or pleural and abdominal effusion may be present. Feline patients with severe AV canal defects generally die due to secondary left heart failure [7]. This patient was presented with severe left-sided and right-sided heart failure that improved after medical therapy. The authors have observed hind limb paresis as a common sign of heart disease in ferrets, which is unrelated to arterial thromboembolism but with limb weakness [19]. This ferret was free from clinical symptoms for at least six months after diagnosis. According to our clinical experience, patients with left-to-right shunting clinically improve or, at least, do not worsen after beta-blocker therapy when titrated gradually.

A diagnosis of AVCD is performed based on cardiovascular examination using imaging methods like radiography, ECG, and echocardiography. Generally, radiography reveals cardiomegaly, especially regarding the right atrium, both ventricles, and a pronounced pulmonary artery [4, 14]. The ECG commonly shows QRS changes compatible with conduction abnormalities, especially bundle branch blocks [3]. However, in the presented case, observed changes were related to the left atrial enlargement (prolonged P waves) probably due to volume overload in the single atrium and increased pressure in pulmonary veins associated with clinical signs of left-sided heart failure. Two-dimensional echocardiography can reveal the presence and size of ASD and VSD and allows the assessment of the AV morphology and the papillary muscles architecture [1]. Although this ferret had several malformations, the left ventricle function was sufficient to provide normal blood pressure and peripheral perfusion. The presence of simultaneous ASD, VSD, tricuspid valve changes and AV insufficiency helped to differentiate complete AVCD from other types of AVCD, common atrium and incomplete ASD associated with mitral and tricuspid dysplasia [4]. Severe systolic pulmonary hypertension was determined by right ventricular hypertrophy and high-velocity tricuspid regurgitation with a peak velocity of 4.39 m/s (82 mm Hg, assuming a right atrial systolic pressure of five mm Hg) that certainly contributed to the complexity of the clinical signs.

Medical management is aimed at slow cardiac remodeling, preserving ventricular function and controlling signs of congestive heart failure with ACE inhibitors, beta-blockers, spironolactone, diuretics, exercise restriction, oxygen, and supportive therapy. In our case, early diuretic, inotropic and vasodilatory therapy significantly reduced left and right ventricular overload and postponed disease progression. Open cardiac surgery for AVCD using extracorporeal circulation has been described in several case reports in dogs and cats [11–13]. Due to the relatively small body size of the patient, and also the high price of the procedure, the owner declined this alternative.

In conclusion, a complete AVCD characterized by a large ostium primum ASD, VSD showing left-to-right shunting and AV dysplasia in a ferret showed clinical signs of heart failure according to history and clinical signs. The final diagnosis was achieved by echocardiography that demonstrated the complex lesions. The ferret survived at least 6 months with only medical treatment suggesting that this approach could be considered in other severe heart congenital heart disease in other species.
Abbreviations
AVCD: Atrioventricular canal defect; ASD: Atrial septal defect; AV: Atrioventricular valves; IVS: Ventricular septum; VSD: Ventricular septal defect

Acknowledgements
Authors would like to thank Mr. Robert Westbrook for the English revision of the manuscript.

Authors’ contributions
All authors have read and approved the manuscript. CA: writing manuscript, echocardiographic interpretation. VJ: patient management, data collection. KH: patient management, data collection. MK: study concept, data collection, radiographic interpretation. MC: data collection, ECG interpretation, data analysis

Funding
This work was supported by the Internal Creative Agency of University of Veterinary and Pharmaceutical Sciences Brno (project number FVL/Chva/ITA2019). The role of funding is to financially support end edition and publish charges.

Availability of data and materials
Data is available at the Small Animal Clinic (internal medicine and diagnostic imaging departments) at the University of Veterinary and Pharmaceutical Sciences Brno.

Ethics approval and consent to participate
This paper was approved by the animal welfare and ethics committee of the University of Veterinary and Pharmaceutical Sciences Brno.

Consent for publication
We confirm the owner has given written consent for their pet’s personal or clinical details along with any identifying images to be published in this study.

Competing interests
The authors declare that they have no competing interests.

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Received: 2 November 2020 Accepted: 25 December 2020
Published online: 22 January 2021

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