



NOTE

Wildlife Science

Anomalous origin of the right coronary artery from the pulmonary artery and congenital bicuspid aortic valve in a California sea lion (*Zalophus californianus*)

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ABSTRACT. A 27-year-old captive female California sea lion (*Zalophus californianus*) died suddenly. Necropsy findings showed severe hemopericardium, the right coronary artery arose from the sinus of the pulmonary trunk, and the aortic valve was composed of two semilunar cusps. Coronary artery branches emerging from the epicardium were dilated and tortuous. Pampiniform vascular plexus formation existed along the adventitia from the coronal sulcus to the pulmonary trunk. This is the first report of multiple congenital cardiac malformations with an anomalous origin of the right coronary artery from the pulmonary artery and a bicuspid aortic valve in a marine mammal.

KEY WORDS: anomalous origin of the coronary artery, bicuspid aortic valves, sea lion

There are few reports of congenital cardiac malformation in marine mammals. Furthermore, there are only two reports of aberrant origin or aplasia of the left coronary artery in small cetaceans and seals [21, 22]. In addition, there have only been some reports of congenital heart defects other than aberrant origin of the coronary artery. Transposition of the pulmonary artery and aorta, ventricular septal defects, a hypertrophic right ventricle, aortic dilation, an atrial septal defect, subvalvular pulmonic stenosis, and hypoplasia of the pulmonary artery and mitral valve have been reported in two dolphins [9, 15]. Septal defects or patent ductus arteriosus have often been reported in Phocidae seals [2–6, 8, 17, 20]. However, the present report is the first to describe anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) and bicuspid aortic valve (BAV) in a marine mammal.

A 27-year-old captive female California sea lion (*Zalophus californianus*) was asymptomatic and died suddenly. Necropsy findings showed substantial hematoid fluid and clots in the pericardial cavity. The myocardium was discolored. Both ventricular chambers were mildly dilated, and the apex was round. The caudal vena cava was diffusely dilated. The aorta from the aortic root to the aortic arch was severely dilated up to 7.3 cm (diameter). The right coronary artery formed the pampiniform vascular plexus (about 3 cm long) which existed along the adventitia from the coronal sulcus to the pulmonary trunk (Fig. 1). A large vessel appeared from the pampiniform plexus, reached the coronary sulcus and descended as the subsinuous interventricular branch after passing across the underside of the right auricle (Fig. 2). The anastomotic vessel, indicating the form of the pampiniformis, was seen near the coronary sulcus of the right ventricular free wall. Branches of the both coronary artery and vein distributed on the posterior wall of the heart were dilated and tortuous (Figs. 2 and 3). Then the right coronary artery originated from the sinus of the pulmonary trunk and the pulmonary valve was normally composed of three cusps (Fig. 4). The left coronary artery, which arose from the aortic sinus, passed between the left auricle and pulmonary trunk, reached the coronal sulcus and divided itself into the ramus circumflexus and ramus interventricularis paraconalis (Fig. 1). The circumflex branch and paraconal interventricular branch were each distributed along the coronary sulcus or paraconal interventricular groove (Fig. 1). These vessels descending from the left coronary artery were severely dilated and tortuous (Fig. 3). The aortic valve was composed of a left and right wall side leaflet (Figs. 5 and 6). The left wall side leaflet connected to the septal cusp of the mitral valve by fibrous tissue (Fig. 6), and it included the origin of the left coronary artery at the sinus of Valsalva (Fig. 5). The right wall side leaflet was larger than that on the left side, and it had a wide ulcer (Fig. 5). Both leaflets of the aortic valve were stiffened; the leaflet on the right side had moderate calcification and sclerosis, and it was stiffer than the leaflet on the left side. The leaflets of both atrioventricular valves were thickened; particularly, the parietal cusp of the mitral valve had fibrotic change and doming. The lung had severe subinvolution

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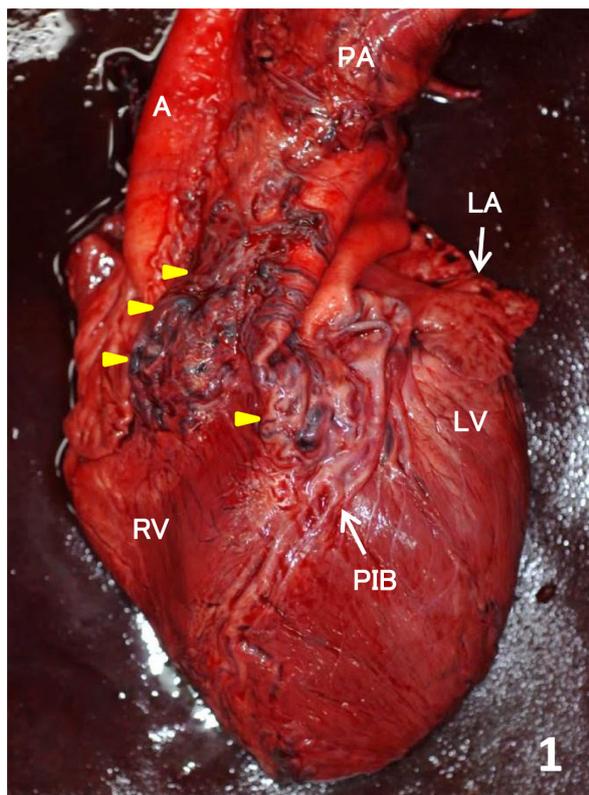


Fig. 1. Photograph showing the entire view of the anterior heart wall. Pampiniform vascular plexus formation (yellow arrowheads) existed along the adventitia from the coronal sulcus to the pulmonary trunk. A: aorta, LA: left atrium, LV: left ventricle, PA: pulmonary artery, PIB: paraconal interventricular branches of the coronary artery, RV: right ventricle.

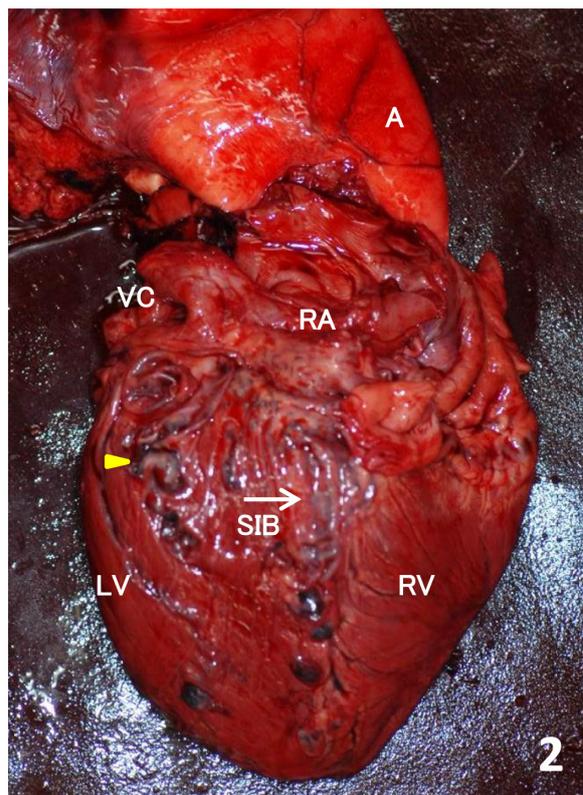


Fig. 2. Photograph showing the entire view of the posterior heart wall. Tortuous and dilated coronary vessels were distributed in the coronal sulcus, subsinuous interventricular groove (yellow arrowhead) and left ventricle free wall. Markedly severe dilation of the aorta was also observed. A: aorta, LV: left ventricle, RA: right atrium, RV: right ventricle, SIB: subsinuous interventricular branch of the coronary artery, VC: vena cava.

and was dark red. Foamy, dark red fluid leaked from the division surface and filled the trachea. We diagnosed the animal as having multiple congenital cardiac malformations, including ARCAPA and BAV.

ARCAPA has been reported in humans and bovine animals [1, 18, 19, 23]. There are a few theories about the cause of an anomalous origin of the coronary artery, including abnormal compartmentation of the coronary orifice by aortopulmonary septum formation of the truncus with abnormal spiral septation, an ectopic coronary artery originating from the coronary orifice and an abnormal connection between the peritruncal vessels and pulmonary artery [1, 23]. The clinical course of a human patient with anomalous origin of the coronary artery from the pulmonary artery is determined by the development of collateral circulation from the normal origin of the coronary artery [12]. Patients with undeveloped collateral vessels develop ischemic cardiomyopathy and die within weeks or months of birth. However, patients with developed collateral vessels survive into adulthood, because the myocardium receives adequate blood supply. When the pulmonary circulation is decreased and retrograde flow of anastomosis occurs, collateral vessels become a left-to-right shunt, and fully oxygenated blood drains into the pulmonary trunk [12]. These patients' clinical presentation varies, ranging from no symptoms to myocardial infarction and sudden cardiac death [18]. BAV has been reported in humans, dogs and horses [11, 13, 16]. The morphogenetic explanation of BAV is fusion of the cusps during embryonic development, and unequal leaflets result in fusion between the two leaflets, causing one leaflet to be larger than the other [11]. Additionally, patients with BAV tend to have calcification of the aortic valve, and they develop significant outflow obstruction, including aortic stenosis and regurgitation over time because of higher mechanical stress [7, 11].

Patients with ARCAPA or BAV may develop complications later, and one's cardiac anomalies can be determined during the antemortem examination. These anomalies have been isolated incidentally by autopsy when patients are asymptomatic or they die suddenly [10, 18].

As the left coronary artery could not supply arterial blood to most of the myocardium, the sea lion examined in the current study may not have survived beyond the early critical period to adulthood. Thus, we suspect that this sea lion had well-established collateral vessel circulation and peculiar anastomosis between the smaller branches for a marine mammal [14, 21]. However, collateral vessel channels may develop into a left-to-right shunt, which causes chronic myocardial ischemia. The pressure



Fig. 3. Magnified view of the ramus circumflexus of left coronary artery in the posterior heart wall. The ramus circumflexus was tortuous and dilated.

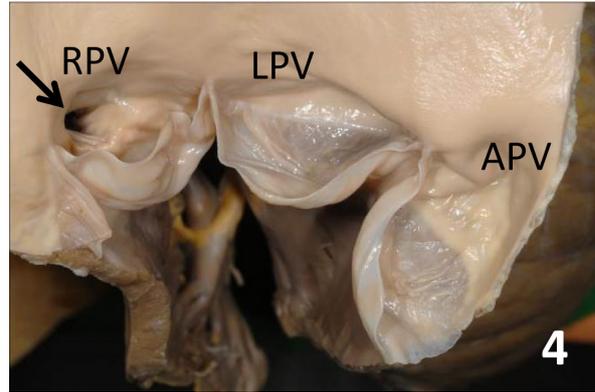


Fig. 4. Pulmonary trunk side view of the pulmonary valve after formalin fixation. The right coronary artery originated from the sinus of the right pulmonary valve (arrow). APV: anterior pulmonary valve, LPV: left pulmonary valve, RPV: right pulmonary valve.

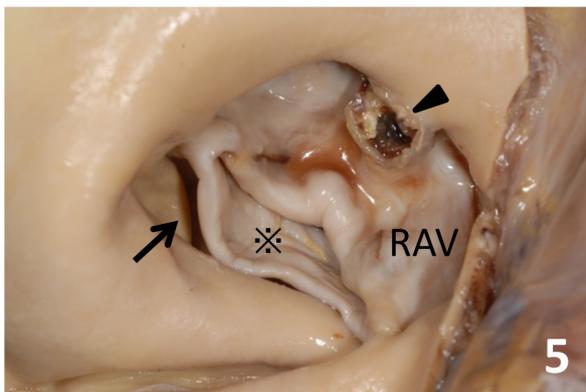


Fig. 5. Aorta side view of the aortic valve after formalin fixation. The aortic valve consisted of two slices that adhered to the right wall and left wall. The right wall side aortic valve (RAV) was severely thickened with sclerosis, and it had a large ulcer (arrowhead) on its base. The left wall side aortic valve that the left coronary cusp and non-coronary cusp fused (⊗) included the origin of the left coronary artery at the sinus of Valsalva (arrow).

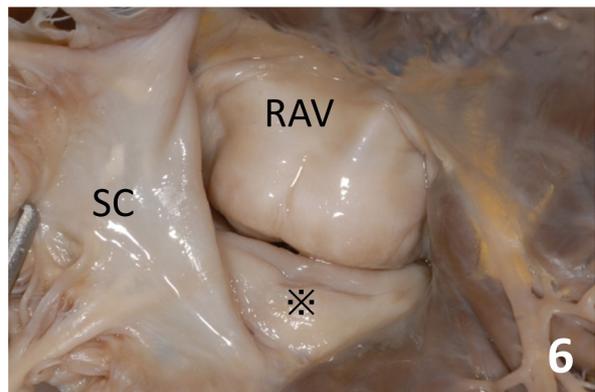


Fig. 6. Photograph of the aortic valve and mitral valve from the left ventricular outflow tract after the formalin fixation. The left wall side aortic valve that the left coronary cusp and non-coronary cusp fused (⊗) was connected to the septal cusp (SC) by fibrous tissue. RAV: right wall side of the aortic valve.

difference and retrograde flow of blood into the shunt can cause the coronary vessels and pampiniform plexus vessels to become dilated and tortuous, and myocardial ischemia may develop because of inadequate blood supply to the myocardium. Chronic

volume overload and ischemia can lead to ventricular dilatation.

The sclerotic valve may have become dysfunctional and caused aortic stenosis or aortic regurgitation. Moreover, the obstruction of outflow, which led to higher mechanical stress, caused fibrotic change and doming of the mitral valve, mild dilation of the ventricular chamber, and severe dilation of the aorta [7, 11]. We speculate that fusion occurred between the left coronary cusp and non-coronary cusp, because the left wall side leaflet was connected to the septal cusp of the mitral valve by fibrous tissue and the left coronary artery arose from the sinus of Valsalva [1, 11].

Although this animal had no symptoms, she may have gradually developed heart ischemia and diastolic failure due to the lesions that caused progressive hemodynamic stress, which increased the risk of sudden death. The animal's clinical history, including the sustained asymptomatic period, sudden cardiac death and pathological lesion produced by a change in the hemodynamic status, is similar to that in previous reports of anomalous origin of the coronary artery from the pulmonary artery and congenital BAV [10, 11, 18, 19, 23]. We consider that the sea lion examined in the present study ultimately developed severe cardiogenic pulmonary edema and died of cardiac tamponade. We suspect that bleeding of the coronal pampiniform vascular plexus and the late complication due to progressive hemodynamic stress caused by ARCAPA and BAV were implicated in cardiac tamponade in this animal.

To our knowledge, this is the first report of ARCAPA and a BAV in a marine mammal.

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