Case #XX

NAME Educational Activities Committee

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Image A – Right atrium and tricuspid valve

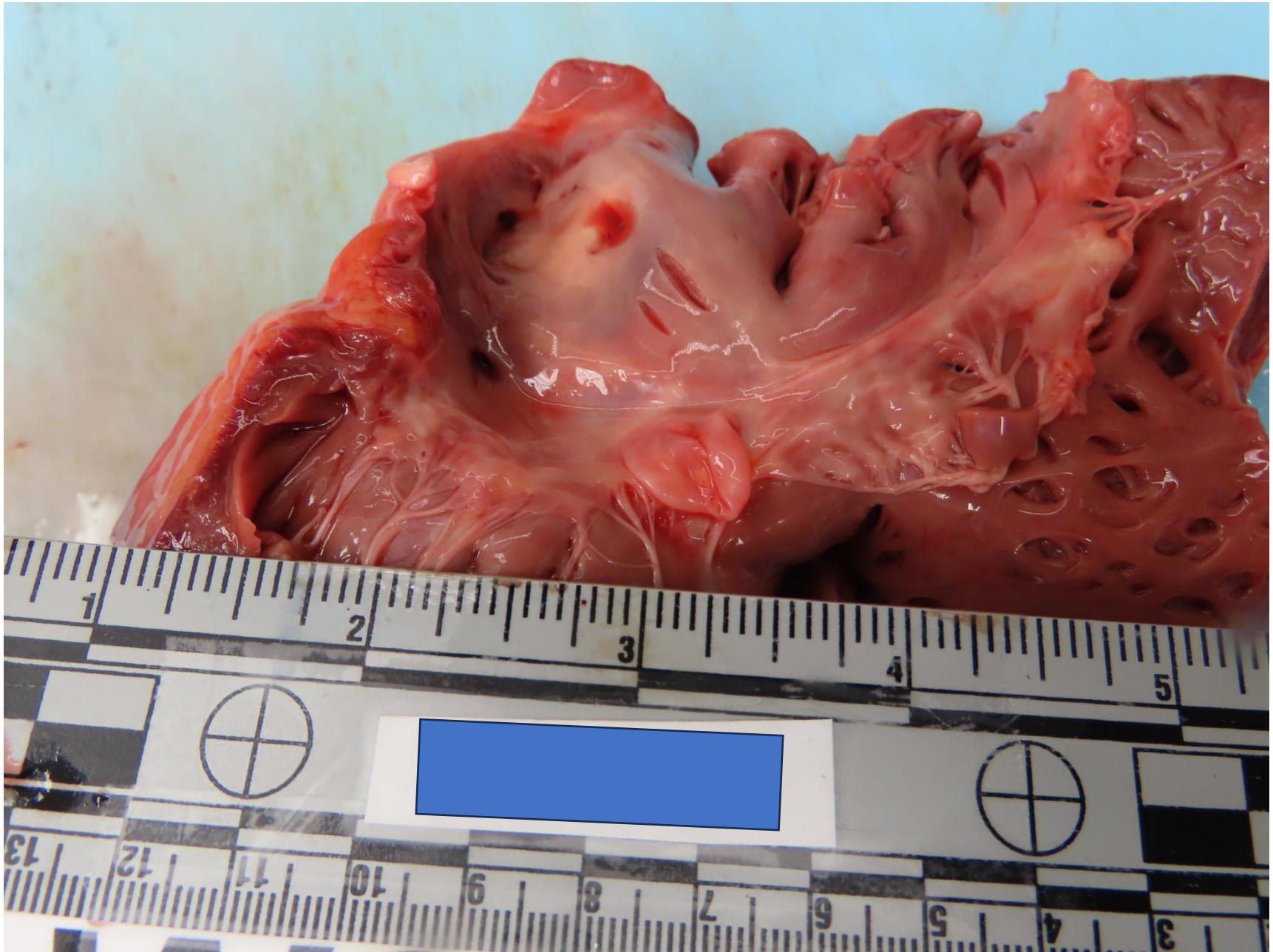


Image B – Right atrium and ventricle

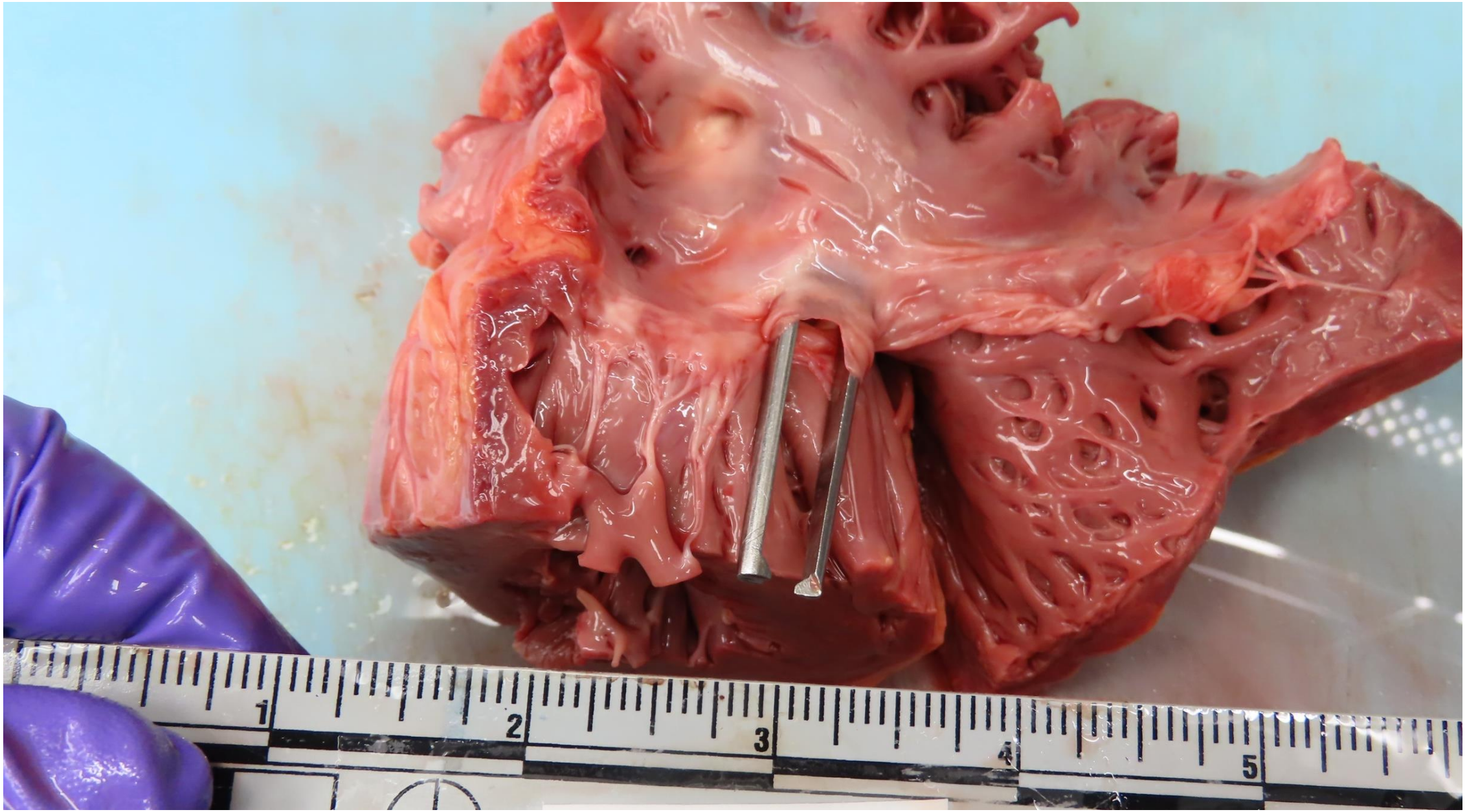
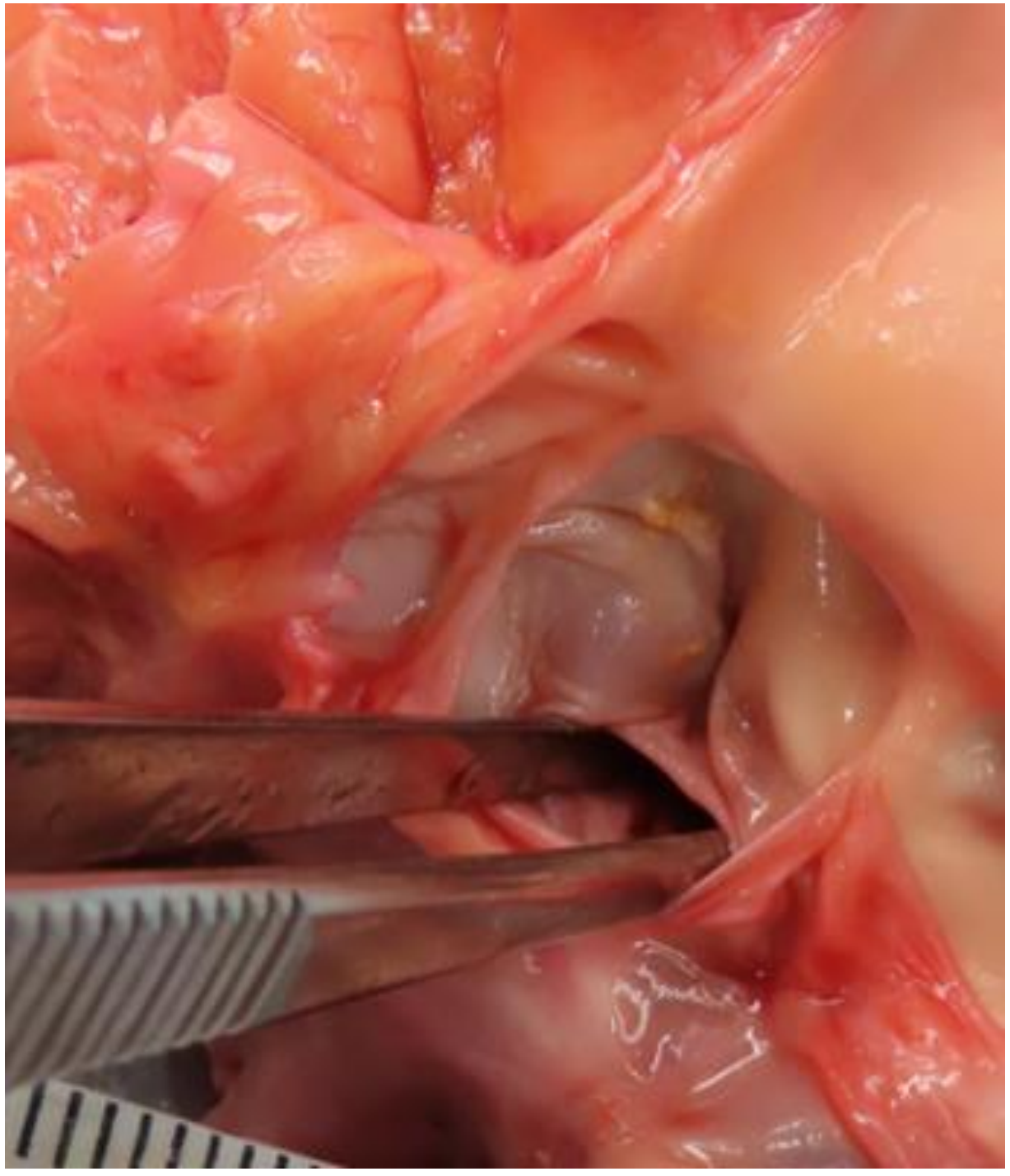


Image C – Aortic valve



A 24 year old man was dropped off at home from work around 1800 hours and later found unresponsive on the front porch at approximately 2300 hours by his roommate when he arrived home. The decedent was found supine with vomit next to him. EMS pronounced him dead on scene. Rigor mortis was present in his legs. He was transported to the Medical Examiner's office for examination. No medical history was available at the time of examination.

Postmortem CT demonstrated cardiomegaly (560 g) and no fractures or intracranial hemorrhages. Autopsy images (A-C) are shown.

Which of the following would be the most likely associated with this autopsy finding?

A. Pulmonary hypertensive changes in lungs
B. Renal artery stenosis
C. Bicuspid aortic valve
D. Blood cultures positive for bacterial sepsis

Answer...

C. Bicuspid Aortic Valve (correct answer: 21.43%)

Images A–C show a ruptured sinus of Valsalva aneurysm (SOVA). A sinus of Valsalva aneurysm is an abnormal focal dilation of one of the aortic sinuses, which are located at the aortic root just above the aortic valve.[1] These aneurysms typically form a saccular outpouching due to weakness of the aortic wall at the junction of the aortic media and annulus fibrosus. The right sinus of Valsalva is most commonly affected, and rupture most frequently occurs into the right atrium or right ventricle, reflecting the close anatomic relationship between these chambers and the aortic root.[2]

SOVAs often remain clinically silent until rupture occurs. When rupture develops, a direct communication forms between the high-pressure aorta and the lower-pressure right-sided cardiac chambers, resulting in an acute left-to-right shunt.[3] Because the pressure gradient persists throughout both systole and diastole, blood flows continuously across the rupture site, leading to sudden volume overload of the right heart and pulmonary circulation. This abrupt hemodynamic change causes rapid deterioration in cardiac function and may result in acute heart failure, malignant arrhythmias, pulmonary edema, stroke, and sudden cardiac death.[1] The prognosis following rupture is poor, with high short-term mortality unless the patient can undergo emergent surgical repair.[1]

Sinus of Valsalva aneurysms are a rare cardiac anomaly and may be either congenital or acquired.[2] Congenital SOVAs arise from an inherent deficiency of elastic and muscular tissue in the aortic wall and are frequently associated with other congenital cardiac abnormalities. Common associations include ventricular septal defects and bicuspid aortic valve, reflecting an underlying developmental weakness of the aortic root.[1] Congenital SOVAs are additionally associated with primary connective tissue disorders characterized by medial degeneration and elastic fiber abnormalities, which further predispose the aortic root to aneurysm formation and rupture.[1]

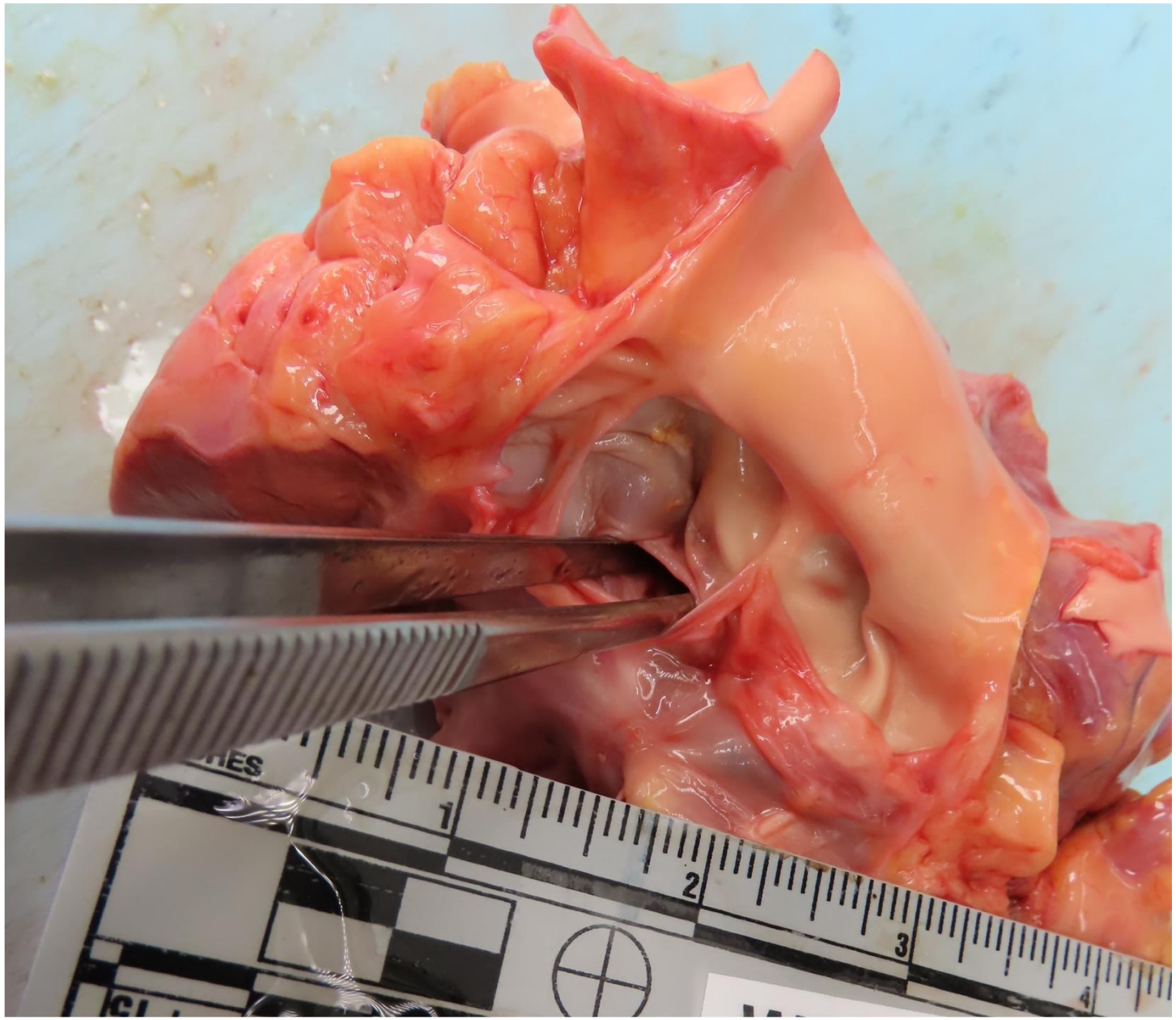
C. Bicuspid Aortic Valve (correct answer: 21.43%)

In this patient, both coarctation of the aorta (shown in photograph D) and a bicuspid aortic valve (photograph C) were present. The co-occurrence of a sinus of Valsalva aneurysm and coarctation of the aorta is extremely rare.[4] Coarctation of the aorta is a congenital narrowing of the aortic arch, most commonly located just distal to the origin of the left subclavian artery.[2] This narrowing produces chronic systemic hypertension proximal to the site of constriction, including elevated pressures within the ascending aorta and aortic root, thereby increasing wall stress and predisposing the already weakened sinus of Valsalva to progressive dilation and eventual rupture.

The presence of a bicuspid aortic valve further increases the risk of aortic root pathology. A bicuspid aortic valve is strongly associated with a congenital aortopathy characterized by medial degeneration, reduced elastic fiber content, and abnormal smooth muscle cell architecture within the ascending aorta and aortic root. These histologic abnormalities weaken the structural integrity of the aortic wall independent of valve function. In addition, bicuspid aortic valves frequently cause abnormal flow patterns and eccentric systolic jets, which increase shear stress on the aortic root and sinuses of Valsalva, further promoting aneurysm formation and rupture.

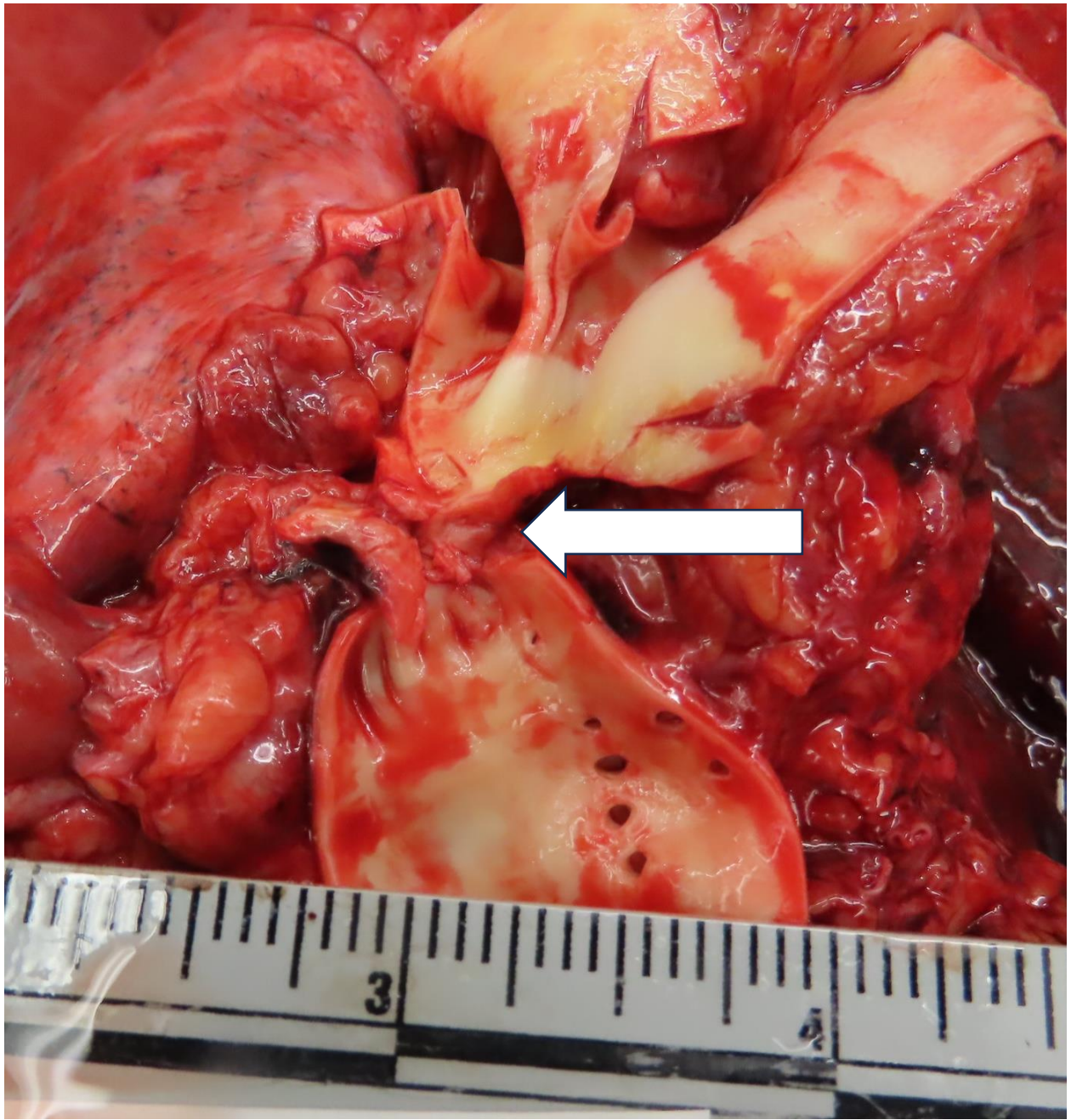
Given the young age of the decedent and the presence of multiple congenital cardiac abnormalities, including a bicuspid aortic valve and coarctation of the aorta, the sinus of Valsalva aneurysm in this case is most consistent with a congenital etiology. Coarctation of the aorta frequently occurs in conjunction with a bicuspid aortic valve, supporting the presence of a generalized congenital aortopathy rather than isolated structural defects.[5] Together, these conditions result in abnormal aortic wall architecture, altered hemodynamics, and chronically elevated proximal pressures. In the setting of a sinus of Valsalva aneurysm, these combined factors markedly increase the risk of aneurysmal rupture and sudden catastrophic cardiovascular collapse. In this case, the presumed mechanism of death is acute heart failure with pulmonary congestion related to sudden rupture of the aneurysm leading to left-to-right shunt.

Image C – Full size



- Bicuspid aortic valve with sinus of Valsalva aneurysm connecting aorta to right atrium

Image D



- Coarctation of the aorta

Other responses…

A. Pulmonary hypertensive changes in the lungs (incorrect answer: 66.14%)

Pulmonary hypertension is defined by increased pressure in the pulmonary arteries, which would also increase the pressure in the right side of the heart.[6] The right side of the heart then has to compensate for this increased pressure, which can eventually lead to right-sided heart failure.[6] Pulmonary hypertension is seen in up to 35% of patients with an atrial septal defect (ASD).[7] Images A-C demonstrate a sinus of Valsalva aneurysm present, not an ASD. The risk for rupture of the sinus of Valsalva aneurysm is increased pressure on the left side of the heart; pulmonary hypertension would not increase the risk of SOVA rupture. In addition, pulmonary hypertension as a complication of SOVA would not typically be seen since the rupture is usually acute (not associated with chronic flow of blood from the left to the right heart).

B. Renal artery stenosis (incorrect answer: 3.11%)

Renal artery stenosis is the narrowing of one or both of the renal arteries and is a leading cause of hypertension in the United States, accounting for 1% to 10% of cases.[8] The most common cause of renal artery stenosis is atherosclerosis, accounting for 60% to 90% of cases.[8] The second most common cause is fibromuscular dysplasia, accounting for 10% to 30% of cases.[8] There are other less common causes, for example, Takayasu arteritis, a systemic inflammation that causes damage to large and medium-sized arteries and their branches.[9] It usually involves the aorta and its major branches, particularly the renal arteries, carotid arteries, and subclavian arteries.[9] There is no evidence of Takayasu arteritis in the photographs. Takayasu arteritis grossly would appear as segmental thickening and fibrosis of the aorta and its major branches, with luminal narrowing or occlusion and a rigid, scarred arterial wall; it is sometimes described as a “tree bark” appearance due to the intimal fibrosis. Additionally, the majority of patients are female and more common in individuals of Asian and Mexican descent.[9]

D. Blood cultures positive for bacterial sepsis (incorrect answer: 9.32%)

Sepsis starts with a systemic inflammatory response that can be characterized by a temperature higher than 38 C or lower than 36 C, tachycardia, tachypnea, or leukocytosis.[10] Then, hypotension develops, and tissue demands are not adequately met with oxygenation. Tissue hypoperfusion can manifest as the end-organ damage associated with septic shock. Hypotension would cause decreased pressure in the chambers of the heart and would not increase the risk of SOVA rupture. Endocarditis can increase the risk of formation and rupture of a SOVA, but there is no evidence of endocarditis in the photographs.[11] Endocarditis grossly would appear as friable, irregular tan-white vegetations on the cardiac valves, often with valvular destruction, ulceration, or perforation, and occasional extension into adjacent myocardium with abscess or fistula formation. Additionally, other infections such as syphilis can increase the risk of development of a SOVA.[12] Although in the setting of both endocarditis and syphilis, these would cause an acquired SOVA.[11,12] Acquired SOVAs tend to present in older adults, whereas congenital SOVAs present in younger individuals.[1] Given the age of our patient, being 24, and other cardiac defects present, this supports a congenital SOVA.

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