# Circle of Willies dependent coronary artery circulation in a neonate with absent left ventricle, aortic valve atresia and type C interrupted aortic arch.

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March 30, 2022

**Abstract**

We present a case of Aortic valve atresia (AVA) with type C interrupted aortic arch (IAA) in a neonate with absent left ventricle and mitral valve atresia. The hemodynamics of the lesion has been discussed with an illustrative cartoon.

A 32weeks old male neonate was delivered vaginally with a birth weight of 1800grams due to premature labour. His right upper and lower limb oxygen saturation was 50%. In view of poor respiratory efforts, he was mechanically ventilated immediately after birth. Chest X-ray revealed bilateral diffuse interstitial reticular pattern of the lungs (Nutmeg lung) suggestive of pulmonary lymphangiectasia. Echocardiography showed intact thick interatrial septum**(Figure 2A)** , atresia of the mitral valve, left ventricle**(Figure 2B)** and aortic valve. The left atrium appeared tense with prominent appendage and the pulmonary veins were dilated**(Figure 2C&D)** . The pulmonary artery arose from the morphological right ventricle(RV) with adequate antegrade flows but the ascending aorta had no connection with the RV, the aortic valve was atretic and the blood flow was retrograde from Circle of Willies**(Figure 2E-F)** since the aortic arch was interrupted after the first branch and the patent ductus arteriosus was supplying the left common carotid artery, left subclavian artery and descending aorta[Type C interrupted aortic arch (IAA)]. There was no aortopulmonary window or visible collaterals from descending aorta. Treatment options like urgent balloon atrial septectomy for survival followed by palliative surgery like Norwood operation or primary heart transplantation were discussed. Considering the guarded long-term outcome, the parents opted for comfort care. A schematic cartoon demonstrating the anatomy and hemodynamics of the described lesion is shown in **Figure 3** .

**Hemodynamics of the described lesion:**

Aortic valve atresia (AVA) with IAA is incompatible with life unless blood flow is provided to the ascending aorta and coronary arteries from bilateral ductus arteriosus or from pulmonary artery through an aortopulmonary window or from descending aorta through collaterals. In the absence of the above source of blood flow, the only possible way for coronary arteries perfusion as illustrated in this case **(Figure 3 A &B)** is through vessels (left carotid and left vertebral artery) arising from the descending aorta flowing to the circle of Willis to perfume the brain and then the hypoplastic aorta and the coronary arteries through retrograde flow via the right carotid artery and right vertebral artery. AVA with IAA type A and AVA with IAA type B can never survive. In the AVA-type B IAA, the only flow to the circle of Willis is the left vertebral artery, which will be insufficient to perfuse together the brain and the coronary arteries.(1) The hemodynamic effects of left side cardiac abnormalities are usually compensated during fetal life, as the fetal circulation allows for the left side of the heart to be bypassed. Once a baby with hypoplastic left heart syndrome(HLHS) takes

its first breath, if the left side structures are inadequate (as in this case mitral valve atresia and absent LV) the pulmonary venous return must shunt across the atrial septum to the right heart, allowing the egress of blood from heart to the lungs via main pulmonary artery and to the systemic circulation via the patent ductus arteriosus. If the atrial septum is intact or restrictive, it leads to critical left atrial hypertension, pulmonary congestion and severe hypoxemia, creating an emergent situation that requires immediate atrial septectomy. This condition will lead to maldevelopment of pulmonary vasculature [lymphatic dilatation, pulmonary venous wall thickening and fibrosis (venous arterialisation)], an appearance termed as nutmeg lung.(2) HLHS with intact atrial septum have the highest risk for mortality. (3) Maldevelopment of the pulmonary vasculature (nutmeg lung appearance) is associated with increased mortality.(4)