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"APVC" - ANOMALOUS PULMONARY VENOUS CONNECTIONS EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Anomalous pulmonary venous connection is an abnormality in the blood flow in which all the 4 pulmonary veins drain into the systemic veins or into the right atrium with or without the pulmonary venous obstruction. The Systemic and the pulmonary venous blood get mix in the right atrium. Becomes an atrial defect or foramen ovale is more important in the left ventricular output as both in the fetal and in the newborn circulations.

Introduction

Anomalous pulmonary venous connection is a rare cyanotic congenital heart defect. Anomalous pulmonary venous connection is shortly called as (APVC).[17] APVC is an abnormality in the blood flow of all the four pulmonary veins that drains into the systemic veins or the right atrium with or without the pulmonary venous obstruction. Anomalous pulmonary venous connection is otherwise called as Anomalous pulmonary venous drainage and Anomalous pulmonary venous return. It is mainly a rare condition of cyanotic congenital heart defect in which all the four pulmonary veins that are malpositioned and make the anomalous connections to the systemic venous circulation.(Normally the pulmonary veins return oxygenated blood from the lungs to the left atrium where it can be then pumped to the rest of the body parts)[16]. A patent foramen ovale, patent duct us arteriosa or an atrial septal defect must be present, or else the condition is fatal due to a lack of systemic blood flow. In some cases, we can able to detect it prenatally.

The APVR can be expressed into four variants, they are Type I: supra cardiac, Type II: cardiac, Type III: infra cardiac, Type IV: mixed pattern. The Supra cardiac (50%), the blood drains into one of the innominate veins (brachiocephalic veins) or the superior vena cava. Cardiac (20%), the blood drains into the coronary sinus or directly into the right atrium. Infra diaphragmatic (20%), the blood drains into the portal or hepatic veins; and a mixed (10%) variant, mixed pattern of any of the above [1,10].

In the case of normal development of pulmonary venous pathways gives us an understanding about the various types of anomalous pulmonary venous return might occur in body. It is due to the failure of the common pulmonary vein to which connect with the pulmonary venous plexus leads to one or more earlier venous connections to the right superior vena cava, and to the left vertical vein, or to the portal vein and the abnormal septation of the sinus venosus can allow the direct connection of the pulmonary veins with the right atrium. Late obstruction of the common pulmonary vein after that the earlier venous channels get disappeared and we can lead to isolated pulmonary vein atresia it is a rare and usually a fatal condition. Failure of incorporation of the common pulmonary vein may lead to stenosis of the common pulmonary vein. All pulmonary venous return connects to the systemic venous system, right atrial and the right ventricular, enlargement occurs, and if any significant pulmonary venous obstruction develops, the right ventricular hypertrophy. The anomalous pulmonary venous connection occurs alone in the two thirds of the patients and it occurs as a part of a group of heart defects (eg, heterotaxy syndromes) in approximately one third of the patients.[2-3]

Pulmonary venous obstruction may occur in all types of anomalous connections, and the clinicians must identify if any sites of obstruction are present or not and should treat the obstruction whenever possible at the time of surgical repair. In supra cardiac connections, the obstruction of vertical vein may occur at its origin, or the vertical vein may be obstructed in the way that it crosses between the left pulmonary artery and the left bronchus. We can able to see certain same In cardiac connections also, in which here the obstruction is to the pulmonary veins seldom that develops but it may occur that the pulmonary venous seldom occurs at the coronary sinus of the common veins junction. Likewise In infra diaphragmatic connections also, there is severe obstruction were the venous flow of pulmonary is almost inhibited with the obstruction of the common pulmonary vein. This obstruction occurs either as it travels through the diaphragm or at its junction with the portal vein system.

Finally in all the types, obstruction may occur because of the restrictive atrial septal defect size and because of small left atrial size.[18 ,19, 20]

This study material aimed through the insight knowledge about the anomalous pulmonary venous connection (APVC). It helps to understand their association with the four pulmonary veins, and various description regarding its development and complications.[4]

Incidence

The incidence of the anomalous pulmonary venous connection (APVC) ranges from 0.6 to 1.2 per 10,000 live births. Among the patients presence with congenital heart disease (CHD), the incidence of APVC ranges between 0.7 to1.5 percent. Anomalous pulmonary venous connection is the fifth most common cause of cyanotic CHD.[5-6]

Ontogenesis for the normal development of Pulmonary veins

In the developing embryo, primordial lungs, larynx, and tracheobronchial tree are derived from a division of the foregut. So, during the early stage of development lungs involves the vascular plexus from the foregut (splanchnic plexus).[22,23,24] At this stage, lungs do not have any direct connection with the heart. There are numerous connections with the splanchnic plexus i.e., umbilico-vitelline and cardinal venous systems. In the early stage of the embryo, the vascular plexus of the foregut involves the lungs buds [25,26,27].

During 27-28 Days a small evagination arises in the posterior wall of the left atrium to the left of the developing septum secundum. It forms the common pulmonary vein. By the end of the first month of gestation, the common pulmonary vein establishes the connection between the pulmonary venous plexus and the Sino-atrial portion of the heart. [11-13] During 32-33 days, the connection between the pulmonary venous plexus and splanchnic venous plexus are still patent. During 38-40 days the connection between the pulmonary venous plexus and splanchnic venous plexus involutes. Common pulmonary vein incorporates into the left atrium in which the individual pulmonary veins connect separately and directly to the left atrium [7-9].

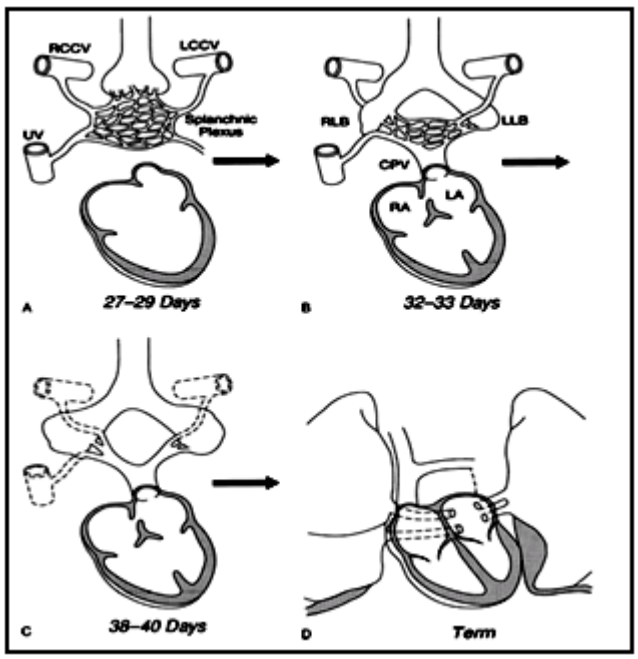

Fig 1: Schematic representation shows the normal developmental series of pulmonary vein and its connections.

Ontogenesis for the abnormal pulmonary venous connections

Normally, during 38-40 days the connection between the pulmonary venous plexus and splanchnic venous plexus involutes. Common pulmonary vein incorporates into the left atrium in which the individual pulmonary veins connect separately and directly to the left atrium.[21,22]

The APVC results in the failure of regression in the established connection between the pulmonary venous plexus and the splanchnic venous system. Sometimes, there is an existence of stenosis in the connection between the common pulmonary vein and left atrium, results in the dilatations of common pulmonary vein called as cor triatriatum sinistrum.

The APVR can be expressed into four variants, they are Type I: supra cardiac, Type II: cardiac, Type III: infra cardiac, Type IV: mixed pattern.[8]

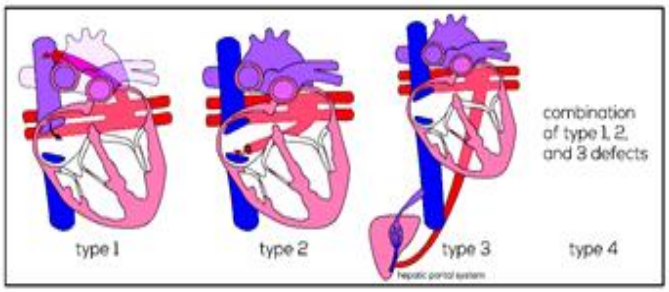

Fig 2: Schematic representation shows the types of abnormal pulmonary venous connections.

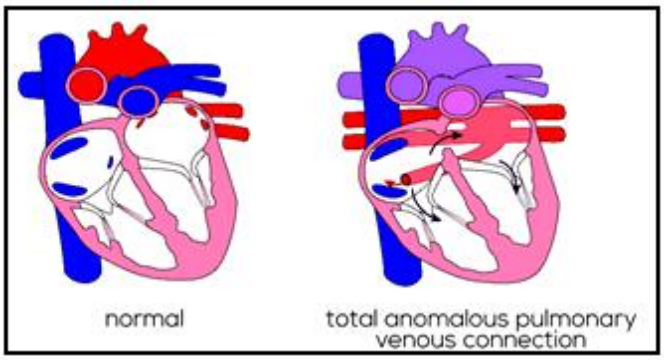

Fig 3: Schematic representation shows the abnormal total pulmonary venous connections.

Discussion

The main aim of this study is to give a major overview of normal pulmonary venous connection, which describes the common variants of the normal and the summarized part of typical patterns of the anomalous pulmonary venous connection. The Pulmonary venous obstruction occurs mainly in all patients with subdiaphragmatic drainage and about 50% of patients with the supracardiac drainage. The Patients with this obstruction develop symptoms early, mainly at the age of 24-36, including with certain conditions of tachypnea, tachycardia, and cyanosis. The signs of the pulmonary hypertension are in progress with the decreasing pulmonary blood flow and worsening cyanosis. Natural history is that of progressive clinical deterioration and the early death in the first week or in the month of life, which all is depending on the degree of pulmonary venous obstruction. Physical findings include severe cyanosis with the significant respiratory distress.. The pulmonary component of the second heart sound is increased and a gallop may be present in this condition. Peripheral pulses are commonly normal after the birth but it may decrease as heart failure progresses also.

Enlargement of the liver commonly occurs, especially in total anomalous pulmonary venous connection.[14]

The Patients with unobstructed pulmonary venous flow present with the symptoms more similar to a very large atrial septal defect. The Often chest radiography in the patients with various respiratory infections reveals the significant cardiac enlargement. Various physical examination findings suggest the right ventricular volume that loading with the increase in right ventricular impulse with a wide split-second sound and pulmonary outflow murmurs with or without a tricuspid diastolic murmur. The Reverse difference in cyanosis has been reported in the newborn period in the total anomalous pulmonary venous connection. In this setting the highly saturated blood in the SVC streams preferentially from right ventricle across the ductus arteriosus to the descending aorta; the lower saturated blood in the inferior vena cava streams across the foramen ovale and flows into the left heart and to the aorta, resulting in higher saturation in the foot than in the right hand[28,29].

Conclusion

This is congenital disorder taking place in the pulmonary venous connection. Anomalous pulmonary venous connection is a rare cyanotic congenital heart defect. Anomalous pulmonary venous connection is shortly called as (APVC). APVC is an abnormality in the blood flow of all the four pulmonary veins that drains into the systemic veins or the right atrium with or without the pulmonary venous obstruction.

In the case of normal development of pulmonary venous pathways gives us an understanding about the various types of anomalous pulmonary venous return might occur in body. The APVC results in the failure of regression in the established connection between the pulmonary venous plexus and the splanchnic venous system. Sometimes, there is an existence of stenosis in the connection between the common pulmonary vein and left atrium, results in the dilatations of common pulmonary vein called as cor triatriatum sinistrum. Surgical treatment can be performed within the first month of life. The operation is performed under the general anesthesia. The four pulmonary veins, which are reconnected to the left atrium, and it is associated heart defects which are mainly surgically closed. With obstruction, the surgery should be undertaken emergently.

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