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"COARCTATION OF AORTA" EMBRYOLOGICAL BASISAND ITS CLINICAL SIGNIFICANCE

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ABSTRACT

The term Coarctation can be defined as the stricture or narrowing of the aorta, it affects both child and adult, but is seen mostly in children. Coarctation can be either is preductal or postductal. Depending on the severity of the condition and the time of diagnosis the treatment can be determined in an individual. Identifying this condition very early in life is also beneficial to the patient even though there would be a few complications that would be faced depending on the form of treatment one choses. The present study aimed to understand the embryological basis and its clinical significance.

Introduction

Coarctation of the aorta may be defined as a constricted aortic segment that comprises localized medial thickening, with some infolding of the medial and superimposed neointimal tissue. The localized constriction may form a shelf like structure with an eccentric opening or may be a membranous curtain like structure with a central or eccentric opening. The coarctation may be discrete, or a long segment of the aorta may be narrowed; the former is more common.

The term Coarctation can be defined as the stricture or narrowing of the aorta [1-3].Therefore, coarctation of aorta is the narrowing of the lumen of the aorta which is the vessel that is responsible for the transportation of oxygenated blood throughout the body which is pumped by the left ventricle of the heart [4].There are two forms of aortic coarctation which are infant form or preductal coarctation and the adult form or postductal coarctation. The ductus arteriosus however, only exist during fetal development and would usually close after birth but instead it remains open or patent hence, it's called patent ductus arteriosus. In this form coarctation obstructs the blood flowing from the heart to the caudal part of the body. This in turn results in the left ventricle having to work much harder in order for blood to be pumped through the narrowed part of the aorta. Thus, there is an increase pressure of blood above the constricted area and lower pressure after the area (constricted area). Aortic coarctation occurs distal to the ligamentum arteriosum. This causes difficulties in the blood flow the same ways as it is in infantile coarctation (high blood pressure in the upper part of the body and low pressure in the lower part of the body). Blood flow also increases in the branches of the aorta which causes high blood pressure in the upper extremities and the head. Due to low blood pressure in the lower part of the body therefore causes patients to have a weak pulse and the lower extremities.

Incidence

The infant form of coarctation accounts for about 70% of cases and it (narrowing/coarctation) occurs after the aortic arch where the common carotid artery, brachiocephalic artery and subclavian artery branches from and before the ductus arteriosus. It is important to note that infantile coarctation only happens during fetal development or can be associated with other congenital changes. It is highly associated with about 5% of infants with turner's syndrome which is a genetic abnormality whereby females have only one (1) X chromosome instead of two (2). The adult form coarctation accounts for 30% of cases and usually develops as an adult. There is no patent ductus arteriosus because it has been closed off and is called the ligamentum arteriosum [5].

Ontogenesis of normal development of the Arch of aorta

The pharyngeal arch develops around the fourth week of fetal development and is supplied by the pharyngeal arch arteries (which are also known as the aortic arch) from aortic sac. [8] The aortic arch has a six paired vascular embryonic structure that gives rise to the great arteries of the head and the neck. The arteries lie anterior to the posterior aorta [8]. In the first pair of pharyngeal arch arteries, most of them would disappear but, the leftover parts of this pair would form part of maxillary arteries and this would supply the ears, muscles of eye, face, and teeth. They (first pair) may also help in the formation of external carotid artery. The posterior part of the second pair of pharyngeal arch arteries goes all the way to form the branches of the stapedial arteries. The stapedial arteries are small vessels that run directly to the loop of the stapes which are small bones in the middle ear.

The proximal part of third pharyngeal arch arteries forms the common carotid arteries which in turn supplies the structures of the head. The distal part of this pair of arteries then unites with the posterior aortae or dorsal aortae to make

up the internal carotid artery which is given the name carotid arch. The internal carotid artery supplies the pituitary gland, brain, meninges (dura mater, arachnoid mater and the pia mater), middle ears and the orbits. The left fourth arch artery of the pharyngeal arch forms a part of the arch of the aorta. The immediate or proximal part of this artery is developed from left horn of the aortic sac while the distal part is formed from the left dorsal aorta. The proximate part of right subclavian artery evolves from the right fourth arch artery, while the right seventh intersegmental artery and the right dorsal aorta form the distant part of the right subclavian artery. It is important to note also that the left subclavian is not a derivative of the pharyngeal arch artery but instead it is developed from the left seventh intersegmental artery [8].

However, for the fifth pair of pharyngeal arch artery their development is not as fruitful as the other four pair because these arteries do not develop in about 50% of persons and for the remaining 50%, only a few capillary loops would develop which would eventually degenerate [8-10].

Lastly, the sixth pharyngeal arch arteries are where the proximal part of the left sixth artery develops and go all the way to form proximal part the left pulmonary artery [6]. A prenatal shunt or the ductus arteriosus is formed when the distal part of this artery is passed from left pulmonary artery to dorsal aorta. The immediate part of the sixth right artery develops into the proximal part of the right pulmonary artery and while the distal part of the sixth right artery regresses [8].

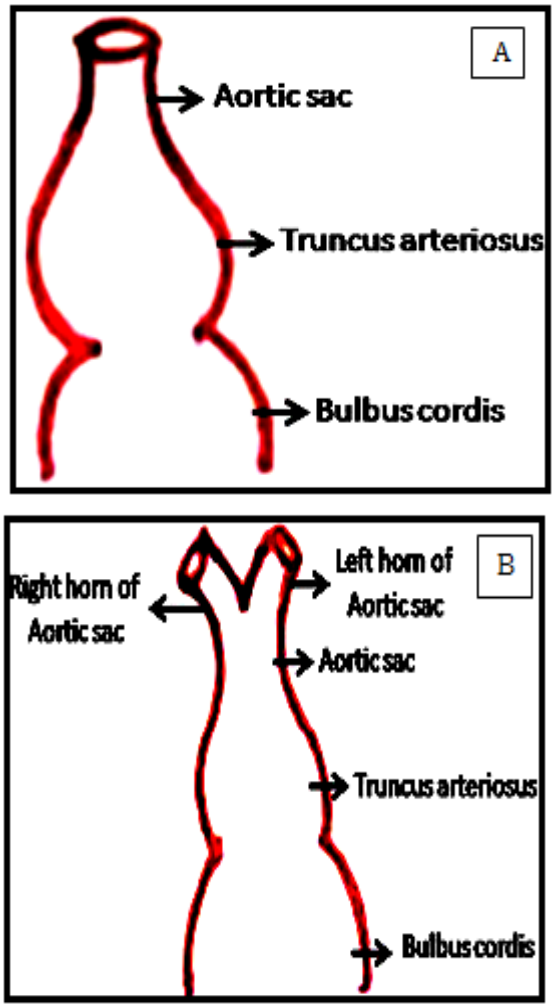

Fig 1: The schematic shows the development of Aortic sacs A. schematics showing the proximal part of the developing heart tube and B. During the later period, the Aortic sac shows its terminal branches called Right and Left horns.

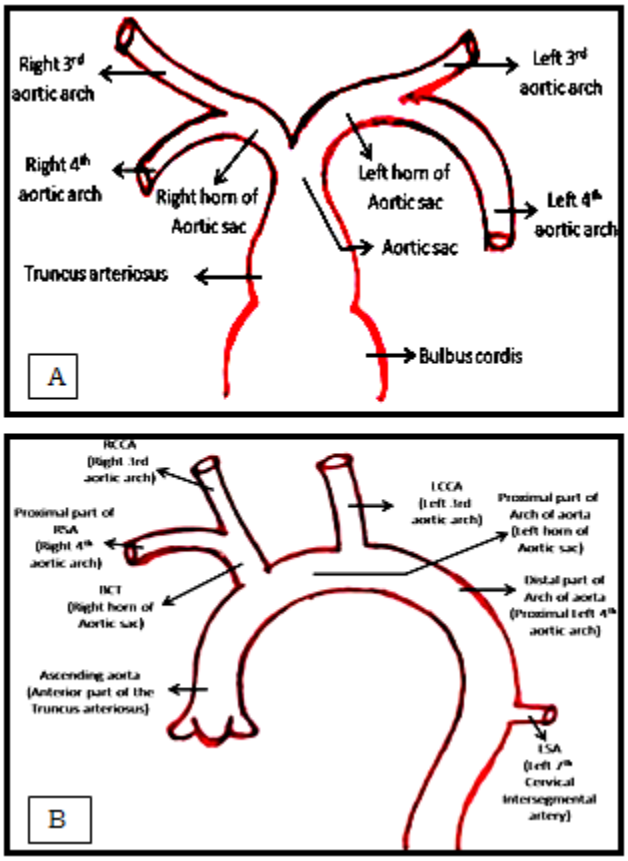

Fig 2: The derivatives of aortic arch arteries A. schematics showing the Truncus arteriosus receives the third (III) and fourth (IV) right and left Aortic arch arteries, which opens into the right and left horns of the Aortic sac and B. Derivatives of the Aortic sac horns and third (III) and fourth (IV) right and left Aortic arch arteries.(BCT- Brachiocephalic trunk, RSA- Right subclavian artery, RCCA- Right Common carotid artery, LCCA- Left Common carotid artery and LSA-Right subclavian artery) [3].

Ontogenesis for the anomalous formation of Coarctation of Aorta

The abnormality of coarctation of the aorta is as a result of the defect in the fourth left and sixth pharyngeal arch arteries. This can be explained by two (2) theories. These theories are the theory of ductus tissue and hemodynamic theory.

Firstly, the ductus theory is as a result of the smooth muscle cell of the ductus migrating to the periductal aorta. This, in turn, causes the narrowing and constricting of the lumen of the aorta. However, this theory doesn't account for all the cases that involve coarctation.

In the second theory, the hemodynamic theory, it is said that coarctation occurs as a result of reduced blood flow and volume through the aortic arch of fetus and isthmus. The isthmus or aortic isthmus is distal to the part where the left subclavian artery is derived from at area of the ductus arteriosus. The aortic isthmus is known for receiving a relatively low amount of blood flow. Most of the blood flow is from the right ventricle to the ductus arteriosus through to the descending aorta. The ascending aorta and the brachiocephalic arteries are supplied with blood via the left ventricle and also during this supply the aortic isthmus receives a small amount of blood [9-20]. Hence, this results in the anomalous development of arch of the aorta with the preductal or postductal coarctation.

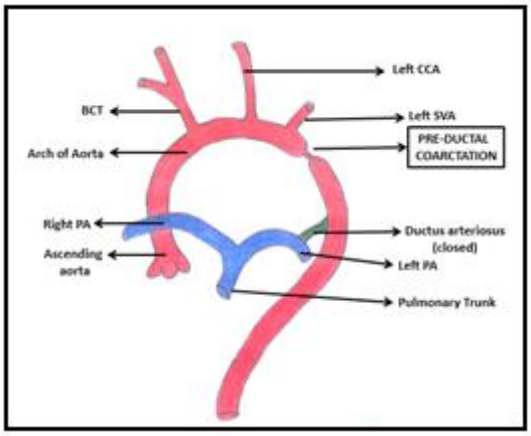

Fig 3: A schematic diagram showing pre-ductal coarctation of the aorta.

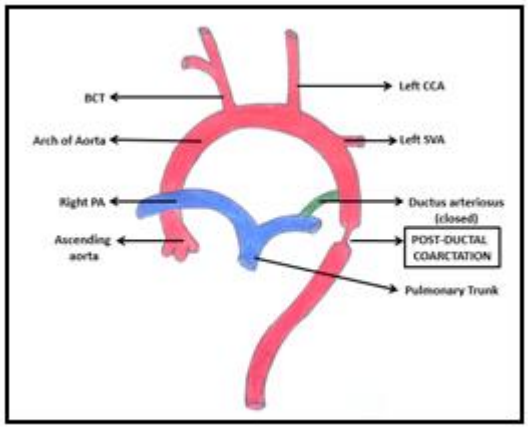

Fig 4: A schematic diagram showing post-ductal coarctation of the aorta.

Discussion

Children with mild cases of aortic coarctation tend to show little to no signs or symptoms in early stages of life. Diagnosing of this condition may not be possible until later in a child's life. Children with coarctation of aorta is said to also have other congenital heart disease such as ventricular septal defect, aortic stenosis and patent ductus arteriosus or mitral valve abnormalities.

In severe cases, after birth babies develop problems due to enough blood not being able to pass through the aorta and to supply the rest of the body. Arterial hypertension is present due to low blood pressure in the lower extremities and a weak pulse is felt from the femoral arteries [5]. The main symptoms are: paleness of the skin, irritation, heavy breathing and poor feeding habit. In infants with poor feeding habit, there is an increase risk in developing other conditions such as necrotizing enterocolitis which is a intestinal disease that occurs in the small or large intestine due to injuries or when the begins to die.

However symptoms may vary depending on the severity of the coarctation. In severe cases of aortic coarctation individuals display symptoms at an early stage in life. Leaving coarctation of aorta untreated in babies may lead to heart failure and death. In the cases of coarctation being diagnosed at a later age in life the most common symptom is hypertension which is also called high blood pressure. Other symptoms for this case may include murmur of the heart, shortness of breath during exercise, weakness of the muscles, cold feet or leg cramps and epistaxis or nosebleeds [20-22].

Aortic Coarctation usually occurs after the left subclavian artery but if it happens to occur before (the subclavian artery), the blood flowing to the left arm would be interrupted which would result in different strengths of radial pulse. There would be a delayed or weak radial pulse in the left arm and normal (radial) pulse in the right arm whereas in the coarctation occurring after the subclavian artery, the radial pulse would be occurring at the same time.

The coarctation of aorta however can be diagnosed accurately with magnetic resonance angiography. Untreated coarctation in adult blood reaches the lower part of the body via the collaterals, for example, the internal thoracic arteries by the subclavian arteries which can be seen on an angiogram. On the other hand an MRI of the chest can be used to identify the location of the coarctation and to conclude if it affects any other blood vessels in the body.

Lastly, cardiac catheterization can also be used in order to measure the real time functioning of the heart. This is done by inserting a hollow plastic tube (introducer sheath) into the arm or leg of the patient. The catheter is then positioned through the blood vessels of the patient until it reaches the coronary arteries. A dye (contrast) is then injected through the catheter's tip while x-ray is being taken in order to follow the dye. This procedure is called coronary angiogram [5].

It doesn't matter the age at which the defect is diagnosed aortic coarctation would need to be fixed one the symptom starts to present. It can either be done with surgery of a procedure known as the balloon angioplasty. This procedure uses a catheter that is inserted into the blood vessels and then it is directed into the aorta. After the catheter had reached the narrowed area in the aorta, a balloon is enlarged in order to expand the blood vessels. In most instances a mesh-covered tube also called stent is use to open the blood vessels.

Surgically, aortic coarctation is corrected by removing the narrow part of the aorta and it is patched in order to allow normal flow of blood through the aorta.

Even though surgery may have been done some persons may still experience high blood pressure but it is treated with medication [19].It is important to keep in mind those patients who may have fixed their coarctation whether by balloon angioplasty or surgically, are not cured from their condition. These patients are required to visit the cardiologist regularly.

Conclusion

It is important for persons with aortic coarctation to seek treatment at an early stage because if left untreated the can have long term effects of high blood pressure due to the coarctation. They can also have other complications such as stroke, rupture of the aorta and coronary artery disease.

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