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"INTERRUPTED AORTIC ARCH" EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

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ABSTRACT

Interrupted aortic arch a rare abnormality, is the discontinuity of the ascending and descending aorta. There are three types which are classified based on the area of interruptions. They are Type-A, Type-B and Type-C. In the Type-A form, the interruptions occurs distal to the origin of the left subclavian artery. Type-B interrupted left aortic arch is characterized by the interruption that occurs between the left subclavian artery and left common carotid artery. Type-C variants are due to the interruption or discontinuity between the innominate artery and the left common carotid artery.

Introduction

The heart is a very important organ of the body and it's responsible for blood pumping. The heart has vessels and channels through which it carries out its task of supplying the body with blood and one of these very important blood vessels is the AORTA [1].

The aorta which is the largest artery in the body allocates blood from the left ventricle of the heart to the rest of the body and it has three divisions: the ascending aorta (where the aorta initially leaves the heart and points toward the head), the arch of the aorta or aortic arch (where the aorta changes direction), and the descending aorta (where the aorta points toward the feet) [2-5].

I will be focusing more on the aortic arch in this article.

Aortic arch is the part of the main artery that curves among the ascending and descending aorta. It leaves the heart and arises, and then inclines back to create the arch.

It also has its own branch which includes the brachiocephalic artery (which divides into right common carotid artery and the right subclavian artery), the left common carotid artery, and the left subclavian artery. These arteries provide blood to both arms and the head [5].

Disruption during the development of AORTIC ARCH can lead to several abnormalities among which is interrupted aortic arch.

Interrupted aortic arch is also known as or abbreviated as IAA. It's the lack of a certain portion in the aortic arch [1].This condition mainly occurs when the aorta is not well or fully developed [6].

According to NCBI, IAA can also be defined as a loss of luminal continuity between the ascending and descending portions of the aorta [2]. Looking into the definitions, we can see that interrupted aortic arch could be a serious condition if not corrected on time surgically after birth because, apart from the arm and head not going to receive proper oxygenated blood, it could also lead to other heart problems such as truncus arteriosus (in which a truncus arteriosus comes out of the right and left ventricles instead of the pulmonary artery and aorta), transposition of the great arteries, aortic stenosis and ventricular septal defect [8].

Incidence

Approximately, the infants with TYPE-A interrupted aortic arch are 30 to 40 percent of all the infants with interrupted aortic arch. The second type which is TYPE-B interrupted aortic arch, accounts for 53 percent of reported cases while TYPE-C interrupted aortic arch accounts only for 4 percent of reported cases. [1]

Ontogenesis for the normal development of the arch of aorta

The aortic arches are a series of combined embryological vascular structures that contributes to several major arteries.

The first pair of aortic arch develops from the curvature of the ventral aorta into the primitive dorsal aorta [8]. This first pair of aortic arch is hidden in the mandibular arch and participates in the formation of the maxillary artery also in the formation of external carotid artery [7].

The second pair of aortic arches appears in the middle of the fourth week.it gives rise to the stapedial and hyoid arteries by crossing the second branchial arches. Nevertheless, the first and second pair of aortic arches degenerates rapidly and is invisible after thirty-one days [9-11].

At the end of the forth week, the third pair of aortic arches becomes visible and gives rise to the common carotid artery in addition with the proximal portions of internal carotid arteries which are associated with the development and supply of the brain.

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The internal carotid arteries are attached secondarily to the cranial portions of dorsal aortas and this forms the remaining [12]. The internal carotid arteries are attached secondarily to the cranial portions of dorsal aortas and this forms the remaining carotid artery while the origin of the external carotid arteries develop from second pair of aortic arch [13-16].

Also, the fourth pair of aortic arches develops at the end of fourth week but it appears shortly after the third arches. They have different development for the right and left sides [16].

The right side of the fourth pair of aortic arches forms the proximal portion of the right subclavian artery and is continuous with the seventh segmental artery. In this phase, the right primitive dorsal aorta disappears while the distal portion of the subclavian artery develops from the right dorsal aorta and the right seventh intersegmental artery [17].

The left side development on the other way round continues as the arch of the aorta which grows considerably and is continuous with the primitive left dorsal artery. The seventh segmental artery or left subclavian artery ascends directly from the aorta [18-20].

The brachiocephalic arterial trunk and the first portion of the aortic arch is formed by the short portion of the right primitive ventral aorta which persists between the fourth and sixth pair of aortic arches [21].

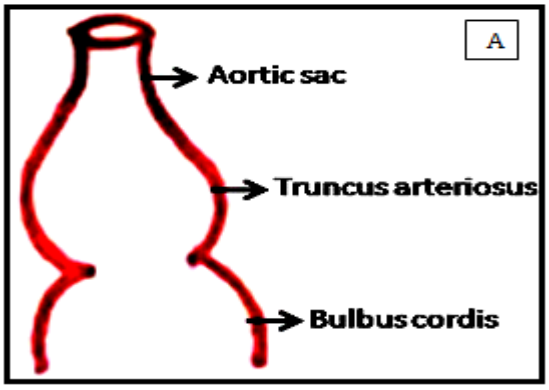
The fifth pair of aortic arches may never develop but in fifty percent of embryos, these arches are undeveloped vessels that degenerate with no derivatives according to research.

The last pair of aortic arches which is the sixth pair, appears in the middle of fifth week and give rise to the right and left pulmonary arteries [20]. Regression happens in the communication with the corresponding primitive dorsal aorta after pulmonary vascularization is established.

This regression is total and complete on the right side. The proximal portion of the right side forms the proximal part of the right pulmonary artery while its distal portion degenerates [21-23].

On the left arch, the proximal portion continues as the proximal part of the left pulmonary artery. The ductus arteriosus is formed by the distal portion of the left arch in which the communication persists with the dorsal aorta until birth and diverts blood from the pulmonary artery to the aorta closure of the ducts arteriosus take place in the neonatal period while the functional duct develops as the anatomic ligamentum arteriosum. [23]

The development of the distal portions of the pulmonary arteries is from shoots of sixth aortic arches that produce the developing lungs. The pulmonary arteries ascend from the pulmonary trunk after the division of the truncus arteriosus. [23-25]



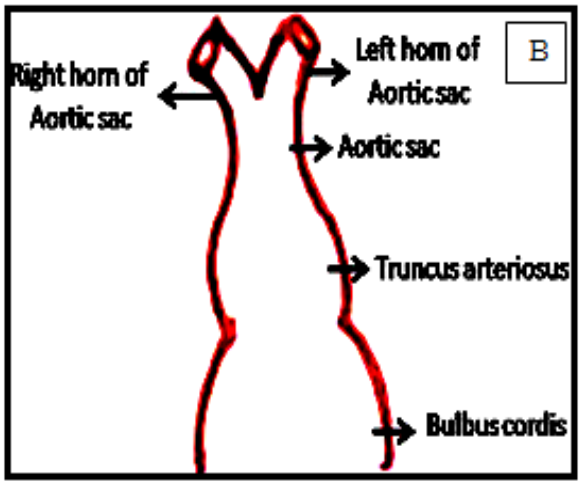
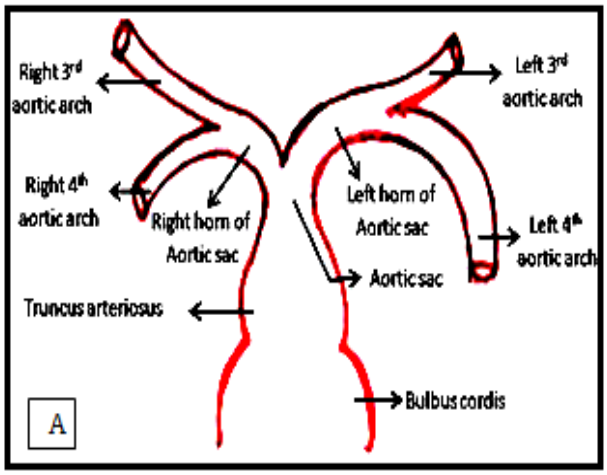


Fig 1. The schematic shows the development of Aortic sacs A. schematics showing the proximal part of the developing heart tube and B. During the later period, the Aortic sac shows its terminal branches called Right and Left horns [3].



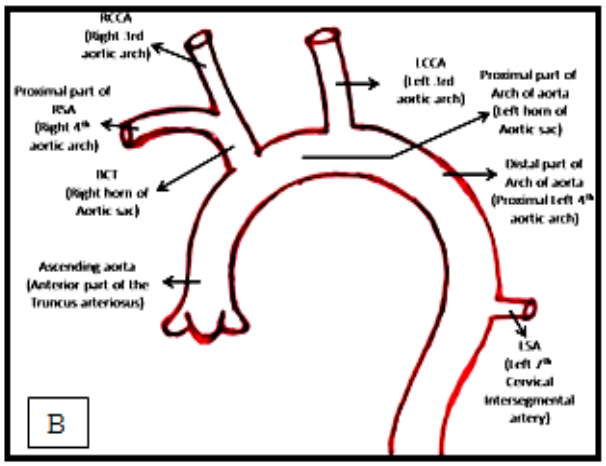


Fig 2. The derivatives of aortic arch arteries A. schematics showing the Truncus arteriosus receives the third (III) and fourth (IV) right and left Aortic arch arteries, which opens into the right and left horns of the Aortic sac and B. Derivatives of the Aortic sac horns and third (III) and fourth (IV) 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].

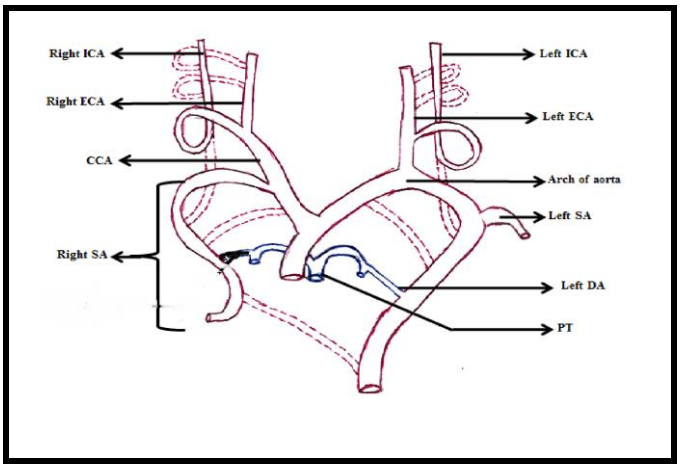


Fig 3.The derivatives of aortic arch arteries. Schematics showing the Truncus arteriosus receives the third (III), fourth (IV) and sixth (XI) right and left Aortic arch arteries. Broken lines are aortic arch arteries that have disappeared. Origin of left subclavian artery is at descending aorta. (ICA- Internal carotid artery, ECA-External carotid artery, CCA-Common carotid artery, SA- Subclavian artery, DA- Ductus arteriosus, PT-Pulmonary trunk).

Ontogenesis for the abnormal Interrupted arch of aorta

Around one half of patients with interrupted aortic arch have a hemi-azygous deletion of a 1.5-3 Mb region of chromosome band 22q11.2 which is the most common deletion syndrome in humans [30-31]. This deletion results in the abnormal development of the heart such as interrupted aortic arch.

In Type-A interrupted aortic arch, this deletion causes the interruption to occur distal to the left subclavian artery which means that the left subclavian artery does not ascend directly from the aorta during the aortic arch development as in case of the normal development (Fig-4). Instead of the normal continuation of the dorsal of aortic arch with the inclusion of left subclavian artery, it continues from the left third aortic arch displacing the left subclavian artery [26-30].

The Type-B interrupted aortic arch occurs between the left common carotid artery and left subclavian artery also due to this deletion because the aorta is not continuous between them (Fig-5). This also implies that the distal part of the arch of aorta, proximal left 4th aortic arch to be precise degenerates which causes gap between the left common carotid and left subclavian arteries [30].

Looking at this type of anomaly Fig-5, the left subclavian artery is not joined to the arch of aorta leaving the innominate and left carotid arteries on the arch of aorta. Since the left subclavian artery is not connected to the aortic arch, it will not be able to receive blood from the aorta talk less of supplying blood to the left arm.

This deletion also occur in Type-C interrupted aortic arch which is between the in-nominate and left common carotid artery. Here, the in-nominate artery does not have connection with the aorta (Fig-6) [30].

In this case, the left horn of aortic sac degenerates. Its degeneration brings interruption between the left common carotid artery and the innominate artery. Since the innominate artery is not connected to the arch of aorta, blood supply to through the right subclavian and right common carotid arteries which are branches of innominate artery to the arm, head and neck will be impossible [31].

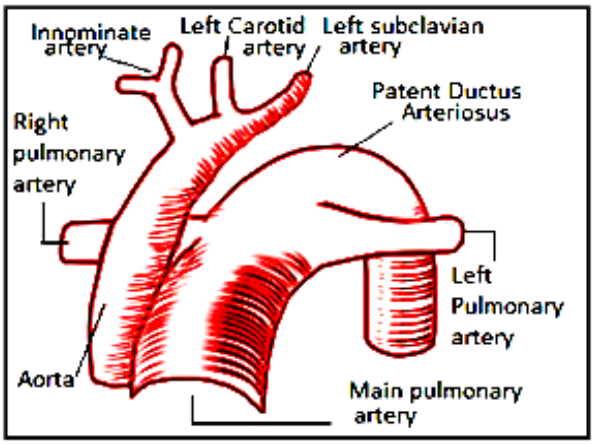


Fig4.Type-A interrupted aortic arch. The interruption occurs distal to the subclavian artery.

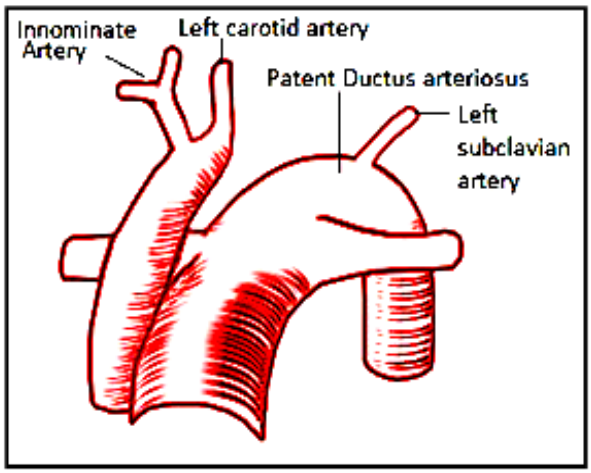


Fig 5. Type-B interrupted aortic arch. The interruption occurs between the left common carotid and left subclavian arteries.

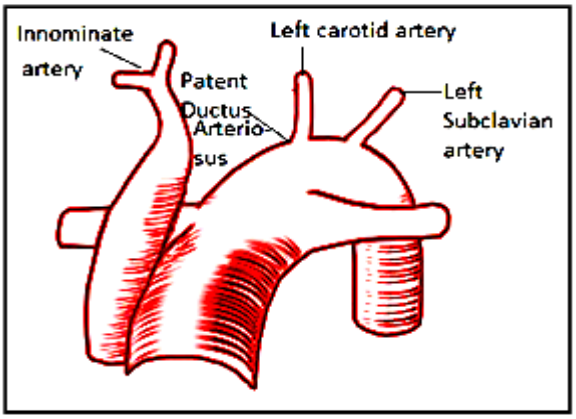


Fig6.Type-C interrupted aortic arch. The interruption occurs between the between the innominate and left common carotid arteries.

Discussion

Interrupted aortic arch comprises about 1% of all innate heart defects. It regularly occurs with other heart defect such as patent ductus arteriosus, ventricular septal defect, transposition of great arteries and aortic stenosis.

Patent ductus arteriosus also known as PDA is often associated with patients of interrupted aortic arch. Patent Ductus Arteriosus is when the ductus arteriosus remains open after third or fourth day of birth which is abnormal. The reason for this opening could be due to less oxygen in the blood of the infant or less oxygen inhaled by the infant after birth [30].

It could also occur due to more prostaglandin in the blood which is derived from the placenta. Another factor is maternal rubella infection during early pregnancy [31].

In a new born baby with interrupted aortic arch, blood flow to the lower part of the body is possible because of the open ductus arteriosus. Blood flow through the 'Ductus Arteriosus' into the Descending Aorta then to the lower part of the body. However, the ductus arteriosus closes gradually after birth and its closure makes the blood pressure in the lower circulation to be inadequate and thence, severe symptoms develop such as difficulty in breathing and impaired kidney function in the first week of life which will call for an urgent surgery [29].

The surgery that will be performed is to reconstruct the aorta and close the ductus arteriosus so that the heart can function normally. Almost all the patients who undergo this surgery survive but when the surgery is not done, it could lead to the death of the infant [30]. Another associated anomaly with interrupted aortic arch is ventricular septal defect also known as VSD. It is the presence of a hole in the septum between the heart's lower chambers (ventricles). This abnormality causes oxygen rich blood to mix with oxygen poor blood because of the presence of the holes.

Ventricular septal defect is associated with interrupted aortic arch because it serves as a shunt that allows blood into the pulmonary artery while Patent Ductus Arteriosus allows the partially oxygenated blood to travel from the pulmonary artery to the descending aorta.Transposition of great arteries is also an anomaly associated with interrupted aortic arch. It is also called transposition of the great vessels sometimes [32].

In this anomaly, the aorta and the pulmonary artery are connected to the wrong ventricles each. In this case, the aorta is connected to the right ventricle which makes the venous blood to be carried to the body instead of the lungs while the pulmonary artery is connected to the left ventricle which makes oxygen rich blood to be conceded back to the lungs instead of the body. This anomaly is associated with Interrupted aortic arch because when there's an interruption in the aortic arches, it could make the aorta o be connected to the right ventricle instead of the left ventricle [33].

Aortic stenosis is another anomaly associated with Interrupted aortic arch. Stenosis means narrowing. In aortic stenosis, the aortic valve is narrowed. The narrowing of the aortic valve results in the hard pumping of the left ventricle to be able to take blood from the blockage of the aorta to the body.

A stenotic aortic valve may have bicuspid valve or unicuspid valve instead of tricuspid valve which is the normal aortic valve. The signs and symptoms of interrupted aortic arch occur when the ductus arteriosus begins to close. This occurs three to four days after birth. The symptoms develop during the first week of life and they include rapid breathing, clammy sweating and poor feeding [33]. The signs can be seen through physical findings or examination. It includes hearing of heart murmur, increase heart rate and breathing rate, absence of pulses on the left arm or leg and possible enlargement of the liver.

Nevertheless, most babies born with interrupted aortic arch have a normal weight and length. So, physical examination is important after birth to know or diagnose if the child has interrupted aortic arch or not.

The evaluation of Interrupted aortic arch can be done with the use of echocardiography, CT, and magnetic resonance imaging (MRI) [34].

Although primary imaging technique for cardiac abnormalities is done by echocardiography, echocardiography is not the best method to evaluate the aortic arch and descending aorta. The best evaluation can be done by the combination of echocardiography and MRI or CT which will enable absolute diagnosis of interrupted aortic arch and related cardiac abnormalities in our patient [5, 6, 8].

Treatment of interrupted aortic arch should be done immediately after it is diagnosed to save the life of the patient.

One of the treatments is to give the patient prostaglandin as soon as possible after birth to keep the ductus arteriosus open before a surgery is performed. When the ductus arteriosus is open with the help of this prostaglandin, it complements blood flow to the descending aorta which will then take blood to the lower part of the body [35].

Surgery is done as soon as possible after this treatment. The surgery is normally done in two stages.

The first stage is to join the two ends of the aorta together while the second stage is to tie off the patent ductus arteriosus which was kept open due to the introduction of prostaglandin to the blood of the infant. Although, overtime, there can be development of stenosis at the site of repair, that may be treated with balloon angioplasty while some needs re-operation such as sub-aortic stenosis [33].

According to research, the outcomes for children with interrupted aortic arch who underwent surgery have greatly improved with 85% to 90% survival. [12]

Interrupted aortic arch can be life saving and not.

It can be life saving in the sense that if the treatment is given and the surgery is done on time, it will help the aortic arch and the heart generally to function normally. Also, blood to the lower part of the body will now be possible after the surgery is done. This will make the patient to live a normal life.

However, if the patient is not given the treatment or if the patient is not allowed to undergo surgery, the patient will not survive because blood will not be able to flow to the lower part of the body which can lead to shock thereby, losing the patient [34].

In addition, children who have interrupted aortic arch have high possibility in getting subacute bacterial endocarditis which is also known as SBE. It is an infection of the heart caused by bacteria in the blood stream.This can arise as a result of dental work or medical measures on the gastro-intestinal tract or respiratory tract because these measures always result in some bacteria entering the blood. Subacute Bacterial Endocarditis can be prevented by taking an antibiotic before these measures.

Children with interrupted aortic arch should not be allowed to do vigorous or competitive exercises or sports. However, they can be allowed to do recreational exercises but they have to be well monitored so that they will be allowed to self- limit their level of exertion [35].

Interrupted aortic arch can be rarely present in adulthood if considerable indemnity circulation develops. Also, the mortality rate of patients with untreated interrupted aortic arch can go above 90% from birth to the first year.

The anomaly can be rarely present in adulthood if considerable indemnity circulation develops. Also, the mortality rate of patients with untreated interrupted aortic arch can go above 90% from birth to the first year. The evaluation of Interrupted aortic arch can be done with the use of echocardiography, CT, and magnetic resonance imaging (MRI).

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Conclusion

In conclusion, interrupted aortic arch is rare in case of adult but common in infants. It is classified into three types based on the area of interruption Type-A, Type-B and Type-C interrupted aortic arches [8].

It is also associated with nonrestrictive ventricular septal defect (VSD) and ductus arteriosus or, less commonly, with a large aortopulmonary window or truncus arteriosus.

Interrupted aortic arches can be treated and surgically corrected. Prostaglandin infusion is an immediate treatment for interrupted aortic arches. Other treatments include Intubation Diuretic therapy, Administration of inotropic medications, Monitoring and correction of abnormal blood gases and electrolytes and Administration of nutrition [10].

Nevertheless, the precise and early diagnosis of interrupted aortic arch and its related cardiovascular anomalies is intensely important for the patient's existence [9].

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