

# Aortic Arch interruption

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[Abstract](#)

[Key-words](#)

[Definition/Classification](#)

[Differential diagnosis](#)

[Etiology](#)

[Clinical description](#)

[Diagnostic methods](#)

[Incidence](#)

[Management](#)

[References](#)

## Abstract

*Aortic arch interruption (AAI) is a rare disease with estimated incidence of 0.003 per 1000 live-births, it is characterized by complete lack of anatomical continuity between the transverse aortic arch and the descending thoracic aorta. AAI should be distinguished anatomically from atresia of the aortic arch where continuity between these segments is achieved by an imperforate fibrous strand of various lengths. This condition is usually not familial but there is a well-known association between AAI and the DiGeorge syndrome with del-22q11. Patients with AAI present heart obstructive lesions, with acute cardiovascular collapse or heart failure after spontaneous closure of the ductus arteriosus in the first days of life. Considerable data indicate that AAI reflects intracardiac malformations producing reduced blood flow to the ascending aorta during the fetal life. Although echocardiography can offer an excellent visualization of the majority of aortic arch anomalies, angiocardiography remains the gold standard in the diagnosis of this disease. Management is mainly surgical; it depends on the degree of subaortic obstruction.*

## Key-words

Newborn, Aortic Arch Anomalies, Left Ventricular Outflow Tract Obstruction, Conotruncal Abnormalities.

## Definition/Classification

Aortic arch interruption (AAI) is a rare disease characterized by complete lack of anatomical continuity between the transverse aortic arch and the descending thoracic aorta. The classification of interruption of the aortic arch is based on the site of interruption: distal to the left subclavian artery (type A); between the left common carotid artery and the left subclavian artery (type B) and between the innominate artery and the left common carotid artery (type C). Patients with interruption and a right-sided aortic arch as well as patients with an aberrant subclavian artery are not taken into consideration by this classification.

## Differential diagnosis

AAI should be distinguished anatomically from atresia of the aortic arch where continuity between these segments is achieved by an imperforate

fibrous strand of various lengths, although the hemodynamic and physiologic effects and the treatment may be identical.

## Etiology

This condition is usually not familial but there is a well-known association between interruption of the aortic arch and the [DiGeorge syndrome](#) with del-22q11. Both the DiGeorge syndrome and interruption of the aortic arch are deemed to be due to an abnormal developmental process involving the neural crest. Among patients with DiGeorge syndrome, 5-20% were found to have AAI while 40-50% of all patients with AAI have DiGeorge syndrome. In particular this syndrome is associated in 60-80% of cases with AAI type B.

### Clinical description

Patients with AAI present, like other ductal dependent left-sided, heart obstructive lesions, with acute cardiovascular collapse or heart failure after spontaneous closure of the ductus arteriosus in the first days of life. Considerable data indicate that interruption of the aortic arch reflects intracardiac malformations producing reduced blood flow to the ascending aorta during the fetal life. As a consequence, interruption of the aortic arch rarely occurs in isolation and a ventricular septal defect (VSD) as well as a left ventricular outflow tract obstruction are almost always present. Virtually, all cases of type B interruption (between the carotid and the left subclavian artery) are associated with a conotruncal abnormality in which a conal septal malalignment type VSD causes subaortic obstruction and hypoplasia of the subaortic region. Characteristically the defect is due to posterior malalignment of the infundibular septum that occurs above the VSD and below the aortic valve so as to encroach upon the left ventricular outflow tract and compromise the flow to ascending aorta. However, different types of VSD and/or different types of left ventricular outflow tract obstructions may be present particularly when aortic arch interruption occurs in association with other complex malformations such as [Truncus Arteriosus](#), [Aortopulmonary Septal Defect](#), [Transposition of the Great Arteries](#), [Double-Outlet Right Ventricle](#), and [Univentricular Heart](#).

### Diagnostic methods

Two-dimensional echocardiography is very important in the diagnosis of AAI and the majority of patients with such a disease may be operated upon only on non-invasive data. The diagnosis should be suspected from the marked discrepancy in size between the ascending aorta and main pulmonary artery, in the presence of the typical malalignment type VSD with posterior deviation of the infundibular septum. Usually, the morphology of the aortic arch and of the aortic branches can be nicely visualized from the suprasternal approach, providing therefore all the information needed. However, in some cases echocardiography may not provide all the details necessary for surgery and angiocardiography remains the method of choice for eliminating possible doubts about the level of obstruction, the distance between the two arch components, associated anomalies, etc. Therefore, even though echocardiography can offer an excellent visualization of the majority of aortic arch anomalies, angiocardiography remains the gold standard in such patients. As any invasive procedure it has some risks that will never be completely eliminated but that compare very favorably against the disasters that can be caused by an inadequate documentation.

### Incidence

AAI has an incidence of 0.003 per 1000 live-births in the New England Regional Infant Cardiac program. At present, there are no data suggesting a difference in fetal prevalence as compared with liveborn prevalence.

### Management

Initial management consists of prostaglandin E1 infusion to achieve ductal patency, with inotropic support if necessary.

The surgical approach to treatment depends on the degree of subaortic obstruction. Diameters of at least 5-6 mm. seem compatible with primary intracardiac repair with VSD patch closure and aortic arch reconstruction. Subaortic regions of 3 mm. or smaller are inadequate to support normal cardiac output in a full-term infant and the subaortic obstruction must be bypassed. This is accomplished by associating the proximal main pulmonary artery with ascending aorta using homograft augmentation to complete the aortic reconstruction, similar to that used for [hypoplastic left heart](#) syndrome (Norwood operation). Pulmonary blood flow is provided by a Gore-Tex shunt if the VSD is left open or by a right ventricle-to-pulmonary artery conduit if an intracardiac baffle from left ventricle to pulmonary artery via the VSD separates the ventricles.

Pulmonary artery banding is not a satisfactory palliation of VSD with interrupted aortic arch since it frequently results in biventricular hypertrophy with progressive subaortic stenosis that eventually complicates definitive repair by any method at a later date.

The aortic arch itself almost always can be reconstructed with direct anastomosis of the two arch components plus homograft augmentation of the reconstructed arch when necessary to achieve adequate arch size. Artificial tube graft connecting proximal and distal aorta should be avoided in the initial operation in infancy because they are rapidly outgrown and complicate primary end-to-end anastomosis at a later date.

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