Management of Interrupted Aortic Arch

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The introduction of prostaglandin E1 in the late 1970s revolutionized the management of interrupted aortic arch. Complete resuscitation should proceed over several days if necessary before surgery is undertaken. One-stage primary neonatal repair with direct arch anastomosis and ventricular septal defect closure is the preferred surgical approach. Selective cerebral perfusion with near–infrared monitoring is being used with increasing frequency. Although repair of interrupted arch is physiologically corrective, it should not be viewed as fully corrective because of the high incidence of important late left ventricular outflow tract obstruction. This may respond to a simple surgical reintervention such as subaortic resection, but in some cases, an extensive procedure to enlarge the left ventricular outflow tract is necessary. However, procedures directed against subaortic stenosis should rarely be used as part of the initial surgical management during the neonatal period. Careful developmental follow-up is needed for all patients because of the high incidence of DiGeorge syndrome, which frequently manifests as moderately severe developmental delay.

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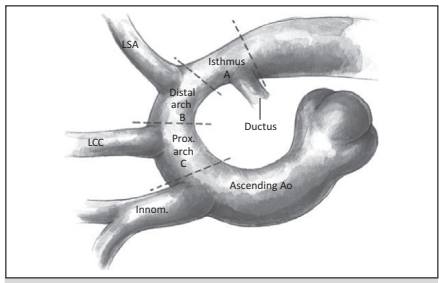

Interrupted aortic arch has been classified by Celoria and Patton.

INTRODUCTION

Interrupted aortic arch is a rare, serious anomaly that carried a high early mortality in the past, but today, it can be managed with the expectation of excellent early survival. However, its association with underdevelopment of the left ventricular outflow tract carries a high risk of need for later surgical reintervention, often on multiple occasions. Furthermore, its association with the DiGeorge syndrome has implications for important developmental delay.

One-stage repair of interrupted aortic arch (IAA) was first described by Barratt-Boyes et al.[1] In the procedure he described in 1972, arch continuity was established using a synthetic conduit. One-stage repair incorporating direct arch anastomosis was first described by Trusler in 1975.[2] Interrupted aortic arch carried an extremely high mortality risk until the introduction of prostaglandin E1 by Elliott et al[3] in 1976. Over the next 5-10 years, it became apparent that careful resuscitation of a neonate, often over a time span of days before proceeding to surgery, was associated with a dramatic improvement in surgical outcome.

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CLASSIFICATION

Using the system of classification devised by Celoria and Patton,[4] the "type B" interruption between the left common carotid and the left subclavian arteries is by far the commonest form of interrupted arch (Fig. 1). In most series, this represents at least 80% of cases. Type B interruption is much more commonly associated with DiGeorge syndrome than type A interruption is, which in many ways can be thought of as an exaggerated coarctation. Type B interruption is often associated with an aberrant origin of the right subclavian artery from the descending aorta. This is important because in this subtype, more flow must pass through the ductus arteriosus during fetal development and less flow through the left ventricular outflow tract and ascending aorta than with the standard type B interruption, that is, with normal origin of the right subclavian from the innominate artery. Thus, it is not surprising to find that the risk of subaortic stenosis is increased when the right subclavian artery arises aberrantly.

Type C interruption occurs between the innominate artery takeoff and the left common carotid. It is extremely rare, having been described in less than 4% of most large clinical and pathologic series.

ASSOCIATED ANOMALIES

A posterior malalignment ventricular septal defect (VSD) is the most common associated anomaly.[5] The posterior

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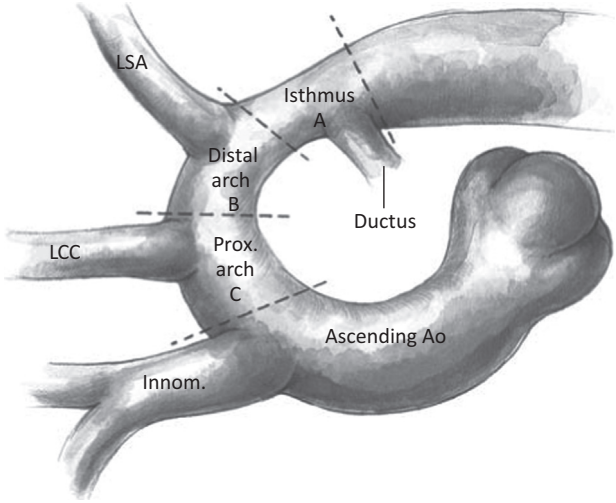

Figure 1. Interrupted aortic arch has been classified by Celoria and Patton as type A: interruption of the isthmus between the left subclavian artery and ductus, type B: interruption of the distal aortic arch between the left common carotid and left subclavian arteries, and type C: interruption of the proximal aortic arch between the innominate artery and left common carotid artery. Ao, aorta; Innom., innominate artery; LCC, left common carotid artery; LSA, left subclavian artery; Prox., proximal.

malalignment of the conal septum relative to the ventricular septum not only results in a VSD but also contributes to left ventricular outflow tract obstruction.[6,7] Other anatomical features that may contribute to left ventricular outflow tract obstruction include the aortic annulus itself, which is usually at least moderately hypoplastic. The aortic valve is frequently bicuspid, and there may be commissural fusion.[8,9] Opposite the septum there may be a prominent muscle bundle on the left ventricular free wall that projects into the outflow tract, the so-called muscle of Moulaert.[10] A fibrous subaortic membrane is almost never seen in a neonate with IAA, but it not uncommonly develops within a year or 2 of repair.[11] An atrial septal defect (ASD) is frequently seen in conjunction with IAA. This is usually in the form of a stretched patent foramen ovale but can be quite large and therefore hemodynamically important.

Other anomalies seen in association with IAA are listed in Table. It can be seen that various forms of single ventricle are seen in 11% of patients with IAA and truncus arteriosus in 10%.

PRENATAL AND POSTNATAL PHYSIOLOGY

Interruption of the aortic arch of any type has no important effect on the fetal circulation. This is not surprising in light of the fact that less than 10% of the fetal cardiac output is usually distributed through the isthmus of the aortic arch.[12] The lower body is perfused prenatally through the ductus arteriosus. Although an abnormal fetal circulation is probably important in contributing to the developmental delay that can be seen with some anomalies such as hypoplastic left heart syndrome and transposition of the great arteries, IAA is one of the congenital cardiac anomalies with the most seriously affected cognitive development. Presumably this is genetic in origin in view of the association between DiGeorge syndrome and developmental delay. It should be noted that neural crest cells play a critical role in aortic arch development as well as central nervous system development.

Following birth, the lower body of the newborn with IAA continues to be adequately perfused so long as the ductus remains patent and pulmonary resistance remains high. Ductal closure in the first day or 2 of life used to be a common reason for presentation in the years before fetal diagnosis was the norm, because it leads to a profound degree of ischemia of the lower body, and in the case of the common type B interruption, the left subclavian artery territory also becomes ischemic.

CLINICAL FEATURES

Prenatal diagnosis by ultrasound is generally made today in more than 50% of patients. The advantage of prenatal diagnosis is that treatment with prostaglandin E1 is started immediately after birth and an acidotic insult is avoided. For a patient who is not diagnosed prenatally with the most common form of IAA, that is, with an associated patent ductus arteriosus and conoventricular VSD, there may be little indication of serious heart disease during the early