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Transverse Aortic Arch Obstruction: When to Go from the Front

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Transverse aortic arch hypoplasia involving some or all segments of the arch (tubular hypoplasia) may exist in association with intra-cardiac anomalies of varying severity. Surgical repair of the distal transverse aortic arch and isthmus are adequately managed by an extended end-to-end coarctation repair in most infants via a left thoracotomy. The surgical management and timing of proximal aortic arch obstruction is controversial but almost always requires an approach via sternotomy using cardiopulmonary bypass.
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Transverse aortic arch hypoplasia involving some or all segments of the arch (tubular hypoplasia) usually occurs in association with intra-cardiac anomalies of varying severity, ranging from a simple ventricular septal defect to hypoplastic left heart syndrome. Uniform narrowing of the whole arch is rare and can coexist with supravalvular aortic stenosis, isolated coarctation, or can occur independently, in both neonates and older patients.[1-3] Surgical repair of the distal transverse aortic arch and isthmus are adequately managed by an extended end-to-end coarctation repair in most infants. The surgical management and timing of proximal aortic arch obstruction is controversial. Although operative results for recurrent coarctation and extensive arch reconstruction in the presence of univentricular circulation (ie, Norwood procedure) have been the subject of numerous publications, only a few small case series have reported outcomes of arch repair in children with hypoplasia of the proximal transverse aortic arch with biventricular hearts.[3-6] The purpose of this report is to review the anatomic, physiologic, and surgical management strategies of transverse aortic arch obstruction to determine when to approach this obstruction from a sternotomy.

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Anatomy

The aortic arch is commonly divided into three segments: (1) proximal, between the bracheocephalic and the left carotid arteries; (2) distal, between the left carotid and the left subclavian arteries; and (3) isthmus, between the left subclavian and the ligamentum or ductus arteriosus (Fig. 1). The anatomic criteria for defining arch hypoplasia is a proximal transverse aortic arch of <60% of the ascending aortic diameter or a distal transverse aortic arch of <50% of the ascending aortic diameter, or an isthmus <40%. One common "rule-of-thumb" for determining transverse aortic arch hypoplasia in neonates is an ECHO internal diameter of <0.1 mm for every 1 kg body weight.

Histologic studies have shown that the hypoplastic segment of the arch has a significantly higher than normal ratio of elastin lamella for the diameter of the vessel, an increase in collagen and a decrease in alpha-actin positive cells that could contribute to a decreased ability of the affected arch to distend.[8] This histologic abnormality appears to have played a part in the pathophysiology of arch obstruction in some of these patients and was particularly evident in the subgroup with isolated arch hypoplasia.

Incidence

The incidence of transverse arch hypoplasia in patients who also have coarctation at the usual juxtaductal region is as high as 30%.[6] Our recently presented review of coarctation demonstrated that 314 (or 31%) of 1,012 patients undergoing initial coarctation repair met the criteria for distal transverse

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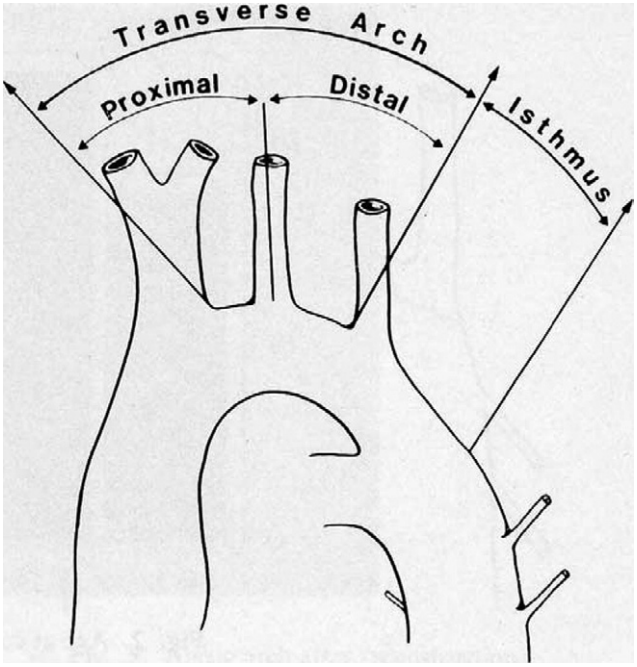

Figure 1 Anatomy of aortic arch. (Reprinted with permission.)

aortic arch or isthmus hypoplasia. Proximal arch hypoplasia requiring subsequent repair, however, was rare in our experience, occurring in only three patients in our 1,012 coarctation patient population.[7] Because of this infrequent requirement for the need to augment the proximal transverse aortic, we usually postpone a surgical approach to the proximal arch and give it time to grow once the distal arch and isthmus obstructions have been corrected via a left thoracotomy and an extended end-to-end or subclavian flap aortoplasty.

Indications for Surgery

The indications for surgical intervention for aortic arch obstruction include symptoms of heart failure, signs of upper extremity systemic hypertension, left ventricular hypertrophy or dilatation, a resting cuff pressure gradient of >20 mm Hg, an exercise cuff gradient of >40 mm Hg, or diastolic continuation of a systolic ECHO gradient.[3] Contemporary clinical studies have shown that the spontaneous growth of the hypoplastic arch commonly occurs in neonates, but in children beyond the neonatal period transverse aortic arch growth is usually inadequate when only the isthmic obstruction is treated.[5]

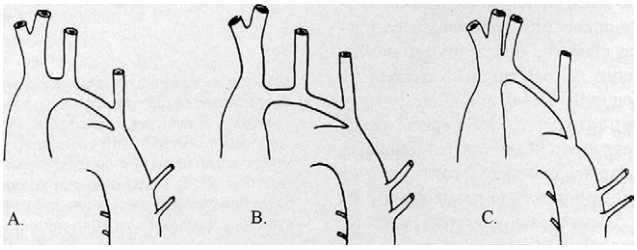

Figure 2 Types of transverse arch hypoplasia: (A) hypoplasia of the distal transverse arch; (B) hypoplasia of both the proximal and the distal transverse arches; (C) absence of the proximal and long-segment hypoplasia of the distal arch. (Reprinted with permission.)

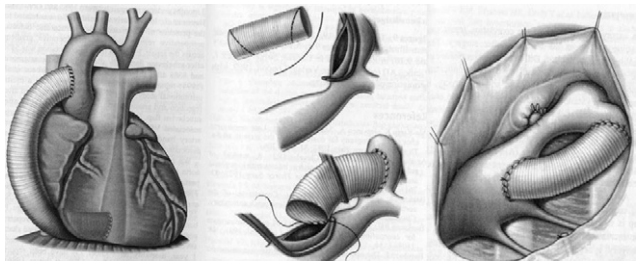

Figure 3 Off-pump extra-anatomic bypass technique. (Reprinted with permission.)

The therapeutic approach to aortic arch hypoplasia is dictated by the location (Fig. 2) and severity of the obstruction, age of the patient, and the associated cardiac lesions.[8]

Therapeutic Techniques

Some of the residual or recurrent lesions of the distal aortic arch and isthmus can be treated by percutaneous balloon dilatation and, in some instances, by implantation of a stent (especially in older children).[9] Even though a mild residual gradient remains in many cases, this approach has been commonly accepted because of the low morbidity it entails. Balloon dilatation, however, has limits, especially in the setting of complex, tortuous lesions or obstruction located in the proximal transverse aortic arch. The optimal surgical approach to these difficult lesions involving the proximal transverse aortic arch is usually an approach via a median sternotomy with (1) hypothermic cardiopulmonary bypass using proximal and sometimes distal perfusion, (2) hypothermia and proximal low flow perfusion, or (3) deep hypothermia and circulatory arrest.

The creation of an extra-anatomic bypass (Fig. 3) between the ascending aorta and the supra-celiac aorta (via sternotomy) or between the ascending and the descending thoracic aorta (using a left thoracotomy) presents the advantages of relative simplicity and efficient relief of the arterial obstruction.[6,10] These approaches, however, show some disadvantages, which in children, might turn out to be detrimental. These grafts have no potential for growth and may lead to grotesque deformities of the aorta with the patient's growth. The development of pseudo-aneurysms at the suture lines can be extremely troublesome and difficult to repair. Aorto-enteric fistula is another potential dire complication that exists with any graft to abdominal aorta anastomosis. Furthermore, any re-operation on the heart (a significant proportion of children have an additional congenital heart defect) will be much more difficult to perform with the presence of a large mediastinal graft. Finally, the loss of elastic capacitance of the aorta can lead to severe left ventricular hypertrophy with its negative long-term effects.

For these reasons, we and other authors have favored an anatomic reconstruction of the aortic arch in children.[5,6] It is our impression that the restitution of an anatomic blood flow

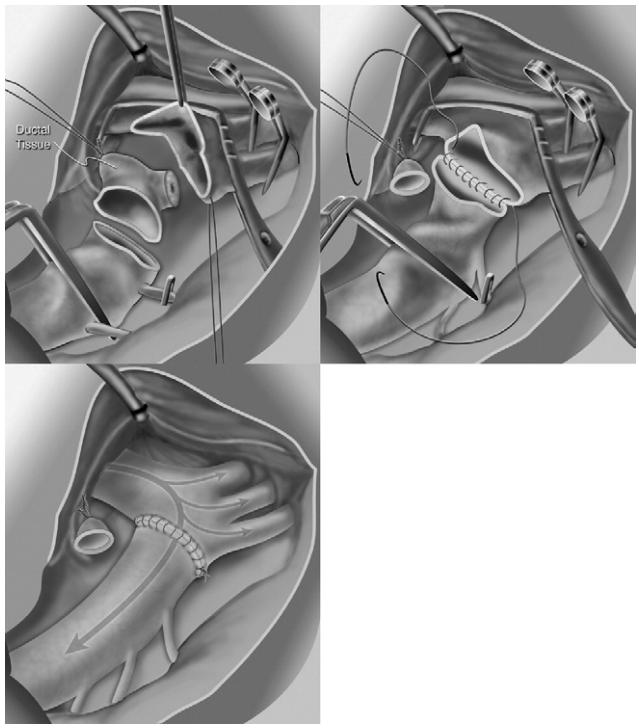

Figure 4 Resection with extended end-to-end anastomosis technique. The aortic arch is divided in the middle of the stenotic segment and an incision is performed in the bottom of the proximal part and in the top of the distal part. Following both segments is re-approximated in a beveled fashion and end-to-end anastomosed with a continuous resorbable suture. (Reprinted with permission.)

in native tissues would provide the best perfusion to the body, the best adaptation during growth, and might preserve the capacitance function of the aorta.

Most proximal transverse aortic arch lesions we have encountered surgically were amenable to repair with three basic techniques:

(1) The sliding plasty (resection with extended end-to-end anastomosis) results basically in the superposition of two stenotic segments with corresponding increase in diameter. This sliding plasty (Fig. 4) is used when the obstruction was relatively long and the tissue shows sufficient elasticity for adequate mobilization during surgery.[11]
(2) Patch plasty is more appropriate for recurrent or persistent lesions, usually in the proximal or middle aortic arch (Fig. 5).[10] For the patch, we have used glutaraldehyde-treated autologous pericardium, Dacron, PTFE, or a cryopreserved pulmonary homograft. Some surgeons advocate use of the adjacent autologous pulmonary artery wall.[6]
(3) Tube graft interposition is reserved for distal long segment arch obstructions in older patients or patients who have developed an aneurysm as a result of a prior surgery or balloon aortoplasty.

Surgery on the proximal aortic arch in the past has involved the use of deep hypothermic circulatory arrest (DHCA; 18°C for less than 35 to 40 minutes). DHCA can be associated with adverse neurological outcomes and in recent years surgeons have focused on decreasing the morbidity associated with aortic surgery and DHCA. A number of centers, including ours, have concentrated efforts to eliminate exposure of neonates and older child to circulatory arrest during complex reconstruction of the aortic arch by: (1) selective cerebral perfusion by cannulation of the innominate artery with or without a Gore-Tex graft attached to the base of the innominate artery; (2) regional low-flow perfusion, by cannulating the innominate artery directly rather than perfusing via a modified Blalock-Taussig shunt (MBTS); (3) continuous low-flow retrograde superior vena cava (SVC) perfusion avoiding both direct shunt and arch vessel cannulation. Cerebral physiologic monitoring with near-infrared spectroscopy has become widely accepted as a cerebral monitoring technique with each of these techniques.[12-15]

Our surgical technique for recurrent or persistent distal arch and isthmus obstruction includes: (1) proximal and distal blood pressure monitoring (keeping the distal blood pressure >40 mm); (2) preoperative and intraoperative somatosensory evoked potential (SSEP) monitoring; (3) redo left thoracotomy (same skin incision) with re-entry of the pleural space one interspace above or below original intercostal space; (4) volume load 10-30 cc/kg to achieve blood pressure proximal to clamp >140 systolic but <200; (5) systemic cooling to 34.5°C [avoid hyperthermia]; (6) left heart bypass or aortic shunt if above conditions encountered; and (7) avoid subadvential dissection plain. When the obstruction involves the proximal transverse aortic arch, we would generally prefer a sternotomy approach using a sliding aortoplasty for virgin cases and a patch aortoplasty for re-do cases.

Conclusion

Incidence of transverse aortic arch obstruction involving the distal arch and isthmus is common in infants and repair utilizing the extended end-to-end repair via a left thoracotomy is safe and yields excellent results. Repair of associated intracardiac lesions can often be delayed with or without placement of a pulmonary artery band. Some centers prefer to treat coarctation with distal arch obstruction associated

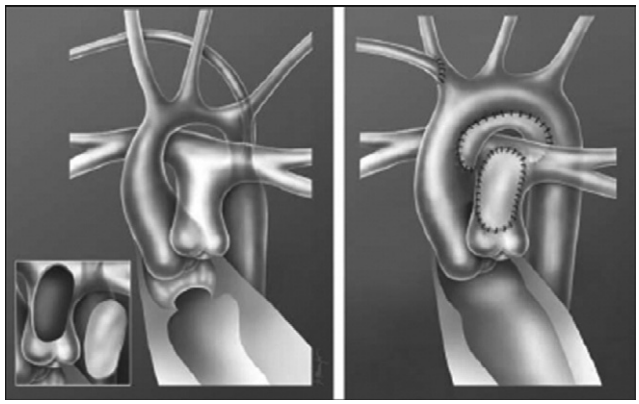

Figure 5 Patch plasty repair. (Reprinted with permission.)

with intracardiac lesions simultaneously and use a sternotomy approach with DHCA to treat both. Significant proximal transverse aortic arch obstruction with or without associated intracardiac lesions is rare, but almost always requires a surgical approach via a sternotomy and most centers correct all lesions simultaneously if possible. A modest increase in mortality and morbidity is often encountered when total correction of both intracardiac and extracardiac pathology is attempted in the newborn period.

Surgical treatment of recurrent or persistent distal transverse aortic arch or isthmus obstruction in older infants and children has greater gradient reduction, less complications, and less recurrence than balloon angioplasty and/or stents insertion. Left heart bypass is rarely needed unless prior balloon angioplasty/stenting has reduced the amount of collateral circulation and/or produced a pseudo-aneurysm. Recurrent or persistent proximal transverse arch obstruction will require a sternotomy, bypass, and one of the several techniques of neuro-protection listed above.

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