

# Transverse Arch Hypoplasia May Predispose Patients to Aneurysm Formation After Patch Repair of Aortic Coarctation

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**Background.** Thoracic aortic aneurysm after patch repair of aortic coarctation is a potentially lethal complication. We hypothesized that transverse arch hypoplasia in association with patch repair of aortic coarctation predisposes aneurysm formation.

**Methods.** A retrospective analysis was performed on all patients undergoing isolated aortic coarctation repair at the University of Virginia Health Systems between 1970 and 1995. Of 244 repairs, 38 patients underwent patch repair. These 38 patients were divided into two groups. The aneurysm group (n = 15) had aortic aneurysms develop at the repair site, which required aneurysmectomy. The nonaneurysm group (n = 23) did not have any aneurysms develop. Univariate analysis and Fisher's exact test were used to identify factors that independently predict aneurysm formation.

**Results.** Univariate analysis demonstrated aortic arch hypoplasia associated with patch repair independently predicts future aneurysm formation ( $p < 0.01$ ). Patients who had an aneurysm develop also had a similar incidence of bicuspid aortic valves, mild restenosis, and late hypertension compared with patients in the nonaneurysm group.

**Conclusions.** Aneurysm formation after patch repair of aortic coarctation develops into a subgroup of patients. Aortic arch hypoplasia associated with coarctation independently predicts future aneurysm formation.

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Vossschulte [1] introduced Dacron patch repair of aortic coarctation (DPA) in 1961. The technical ease and consistent results of this technique were offset by aneurysmal degeneration in a significant proportion of patients. Explanations for this included breaching of the structural integrity of the aorta when the ridge was excised [2], congenital abnormality of the aortic wall [3], scarring when the procedure was done as a redo operation [4], and the persistence of arterial hypertension after the initial repair [5]. Our group [6] previously described the altered hemodynamics across the patch repair site as the advancing wave front is directed toward the native aortic wall opposite the inflexible Dacron patch.

Aneurysm formation due to poststenotic dilatation, a phenomenon of turbulence distal to an obstruction, is a well known pathologic entity. Although all patients undergoing DPA are at risk for aneurysm and require life-long follow-up, we hypothesized that patients with concomitant transverse arch hypoplasia are at an increased risk for aneurysm formation.

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## Material and Methods

Approval for the study was obtained from the Human Investigation Review Committee of the University of Virginia. A retrospective analysis was performed on 244 patients who underwent repair of an isolated coarctation at the University of Virginia between 1975 and 1995. Thirty-eight patients were identified who underwent DPA and were included in this study. The patients were divided into two groups. The aneurysm group (AG) (n = 15) had aortic aneurysms develop at the repair site, requiring aneurysmectomy. The nonaneurysm group (NAG) (n = 23) did not have any aortic aneurysms develop. Aortic aneurysm was defined as the diameter of the repair site greater than or equal to 1.5 times the aortic diameter at the level of the diaphragm determined by echocardiography. Hypoplastic transverse arch was defined as an arch diameter less than or equal to 0.9 times the aortic diameter at the level of the diaphragm. The variables studied are listed in Table 1. Univariate analysis and Fisher's exact test were used to identify factors that independently predict aneurysm formation. Data are reported as mean  $\pm$  standard error of mean. Statistical significance was assigned to values in which  $p$  was less than or equal to 0.05.

Table 1. Aneurysm Group and Nonaneurysm Group Comparisons and Associated Findings

	Aneurysm Group (n = 15)	Nonaneurysm Group (n = 23)	Fisher's Exact Test (p Value)
Bicuspid aortic valve	8 (53%)	7 (30%)	0.397
Late aortic stenosis and aortic regurgitation	5 (33%)	4 (17%)	0.228
Re-stenosis	5 (33%)	6 (17%)	0.449
Mean gradient (mm Hg)	36	30	0.212
Late hypertension	3 (20%)	2 (9%)	0.297
Ascending arch dilatation	4 (27%)	1 (4%)	0.068
Transverse arch hypoplasia	10 (66%)	3 (13%)	0.009

**Results**

The mean patient age at the time of coarctation repair was 22.1 ± 9.1 months in the NAG versus 23.7 ± 8.8 months in the AG (p = 0.89). Gender distribution was similar in the two groups, 80% of the patients who had aneurysms develop in the AG were male versus 70% in the NAG. A bicuspid aortic valve was present in 8 patients in the AG versus 7 in the NAG. Furthermore, 33% of all bicuspid valves in the patients in the AG had moderate to severe regurgitation or stenosis develop, whereas those patients in the NAG had a more benign course with only 17% progressing to a more significant disease, although this did not reach statistical significance. Recurrence of mild gradients over the repair site occurred with similar frequency, (33% and 26% in the AG and NAG, respectively). Three patients had late hypertension develop in the AG and 2 in the NAG. The presence of transverse arch hypoplasia was significantly higher in the AG versus the NAG (8 [53%] vs 3 [13%], respectively; p = 0.01). Linear growth and development was similar in both groups, and there was no significant difference in blood pressure during follow-up between the two groups.

The 15 aneurysm patients in this cohort (Table 2) had surgical repair with aneurysmectomy and interposition graft. The mean elapsed time from coarctation repair to aneurysm repair was 10.2 ± 6.4 years. Access was made through a standard left thoracotomy, and a simple clamp and sew technique was used to insert the tubular graft. No surgery-related mortality occurred. There were two late deaths (1 suicide, 1 trauma-related) many years after the repair. Perioperative morbidity was limited to arterial hypertension; no perioperative hemorrhage occurred and, despite no use of bypass techniques, no neurologic injury occurred. The clamp times were all less than 30 minutes, and all patients appeared to have well-developed collaterals.

**Comment**

Hypoplasia of the transverse arch is commonly seen in infants who have coarctation of the aorta [7]. Hypoplasia is more often present when the coarctation is associated with intracardiac lesions (complex coarctation), which diminishes forward flow to the ascending aorta. Machii and Becker [8] demonstrated a lack of α-actin positive

cells in the aortic wall of hypoplastic transverse arches suggesting limited growth potential. Limited arch growth leads to increased blood-velocity in the arch and turbulence in the descending aorta. Siewers and colleagues [9] demonstrated that in the absence of intracardiac shunts, infants with coarctation and mild arch hypoplasia do well with coarctation repair alone. It is our belief that growth is inadequate in many hypoplastic arches. Pronounced arch obstruction leads to poststenotic dilatation, which has a central role in the pathogenesis of aneurysm formation. Bogaert and colleagues [10] support this belief with magnetic resonance imaging evidence of transverse arch hypoplasia in virtually all patients with aneurysm formation after patch aortoplasty. Most likely, the hemodynamic nature of prosthetic material as previously described by our group [6] adds to this phenomenon with resultant aneurysm formation. Briefly, DPA replaces a portion of the aorta with a tensile strength material that differs from that of the aorta. Although this analysis focused specifically on the use of Dacron, any prosthetic

Table 2. Aneurysm Group Patient Characteristics and Associated Cardiac Defects

Aneurysm Group Patient No.	Age at First Operation	Arch Hypoplasia	Age at Aneurysm Repair	Associated Cardiac Defects
1	2 y	Yes	16 years	None
2	6 y	Yes	22 years	None
3	1 mo	Yes	1 year	VSD, L-TGA
4	1 mo	Yes	16 years	VSD
5	8 s	Yes	18 years	None
6	23 s	Yes	2 years	None
7	20 s	Yes	17 years	VSD
8	1 mo	Yes	4 years	VSD
9	17 s	Yes	8 mo	LSCA distal to coarct
10	3 y	Yes	17 years	None
11	9 y	No	18 years	None
12	3 y	No	16 years	None
13	6 y	No	23 years	Nonse
14	3 mo	No	6 years	VSD
15	1 mo	No	5 years	ASD

ASD = atrial septal defect; LSCA = left subclavian artery; L-TGA = levo transposition of the great vessels; VSD = ventricular septal defect.

material would manifest different tensile strength and compliance than the native aorta. When the aortic pulse wave reaches the stiff, less compliant patch, turbulence is transmitted to the more elastic aorta opposite and adjacent to the patch. This transmitted turbulence subsequently leads to progressive weakening and dilatation of the aortic wall. In addition, as Moulton [11] described, the technique of patch aortoplasty leaves abnormal residual juxtaductal tissue behind, and it is this tissue that is exposed to the abnormal pulse wave opposite the patch. This theory is supported by the hemodynamic and hypertensive characteristics of this cohort. Although there was a trend toward larger trans-repair gradients in the AG group (36 mm Hg vs 30 mm Hg), this trend did not reach statistical significance nor did the incidence of late hypertension (3 vs 2, respectively) (see Table 1). These observations suggest that the postrepair aneurysmal changes were most likely the result of abnormal flow characteristics associated with poststenotic turbulence rather than pressure gradients.

Aneurysm at the repair site after Dacron patch repair of aortic coarctation is a potentially lethal complication and patients who undergo this procedure require life-long follow-up. Our aim was to identify patients at increased risk for having this life-threatening complication develop. Arch hypoplasia in itself is indicative of a more widespread abnormality of the aorta and its branches. Although aneurysm formation may largely be caused by poststenotic dilatation, transverse arch hypoplasia appears to predispose a patient to have other complications of coarctation develop, including dilatory abnormalities of the ascending and descending aorta. Although there were no manifestations of suture line false aneurysms, we found abnormal dilatation is not limited to the repair site in the descending aorta, but can occur in the ascending aorta as well. Five patients in our cohort (4 with hypoplasia, 1 without) had aneurysmal disease of the ascending aorta pointing to a more widespread abnormality in patients with apparently isolated coarctation.

Defects of the aortic wall have been ascribed to mesenchymal abnormalities as well as defects in elements of connective tissue (cystic medial necrosis) [12]. This observation was not confirmed pathologically in our AG cohort and seems an unlikely mechanism in this subgroup of patients. Like most disease processes, the cause of aneurysm formation is complex and often multifactorial. However, our results indicate aortic arch hypoplasia is an independent predictor for aneurysm formation after Dacron patch repair of aortic coarctation. After abandoning DPA techniques we have used subclavian flap aortoplasty intermittently with satisfactory results. In this particular cohort we have repaired one aneurysm (not included in this study) and are currently following another.

The majority of patients who had aneurysms develop in our series were repaired in early adulthood. The progression time in the AG ranged from 1 year to 18 years. Extensive follow-up of our cohort of patched patients demonstrated the initiation of peri-coarctation

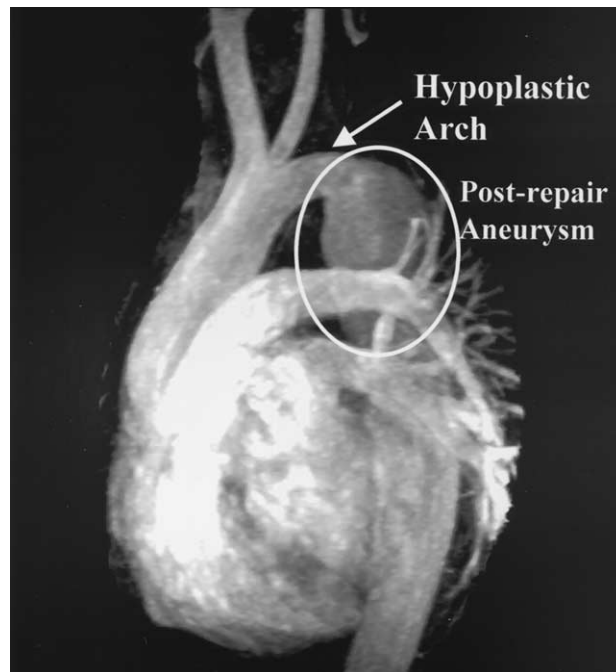


Fig 1. Magnetic resonance angiogram (T2-weighted). Arrow pointing to a 1.2-cm hypoplastic arch ( $> 16$  mm Hg gradient) and a poststenotic aortic aneurysm.

dilatation occurred around puberty in the majority of patients with arch hypoplasia. We propose that accelerated somatic growth and increased cardiac output lead to higher blood velocity, supporting our theory of poststenotic dilatation as an etiologic factor. In our experience, once dilatation at the repair site begins, it is progressive and never remains stable. However, the rate of progression is variable, and frequent imaging is mandatory. Figure 1 is a magnetic resonance angiogram of a 16-year-old boy with previous DPA coarctation repair and hypoplastic aortic arch demonstrating the poststenotic aneurysmal change.

Repair of aortic coarctation using the DPA technique has previously been accepted as an alternative to end-to-end anastomosis. The morbidity and potential mortality of aneurysm development in the peri-coarctation region associated with this technique is well documented and must be considered. In conclusion, we believe that prosthetic patch enlargement of the aorta should be avoided whenever possible as a treatment option for aortic coarctation, particularly in patients with a hypoplastic arch. Patients who undergo this technique of coarctation repair require life-long follow-up. We encourage coarctation resection with extended end to undersurface of aortic arch anastomosis to address both the aortic arch hypoplasia and juxtaductal stenosis. Augmentation with a piece of pulmonary homograft is also a beneficial option. Findings of arch hypoplasia or other obstruction proximal to the repair site are indicative of a predisposition to aneurysm formation mandating these patients be followed closely and frequently.

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