Growth Characteristics of the Aortic Arch After the Norwood Operation

WILLIAM T. MAHLE, MD,*† JACK RYCHIK, MD, FACC,*† PAUL M. WEINBERG, MD, FACC,*† MERYL S. COHEN, MD*†  
Philadelphia, Pennsylvania

Objectives. We sought to characterize the growth of the reconstructed aortic arch after the Norwood operation (NO).

Background. The first stage of surgical palliation of hypoplastic left heart syndrome (HLHS), the NO, includes augmentation of the aortic arch with homograft. Growth characteristics of the reconstructed aortic arch, which is comprised of both native aortic tissue and homograft, have not been characterized.

Methods. Retrospectively, we examined the serial echocardiograms of 50 patients with HLHS who underwent NO to determine the diameter of the reconstructed transverse arch. Measurements were taken immediately after NO and at two other points (1 to 11 years of age). In addition, the autopsy specimens of 10 other patients with HLHS who underwent NO were examined to determine the contribution of native aortic tissue to the overall size of the reconstructed arch at the time of death (12 to 34 months).

Results. The diameter of the transverse aorta increased after NO in all subjects. Its rate of growth paralleled that seen in the normal population, though the reconstructed arch had a significantly larger diameter throughout childhood. Examination of autopsy specimens demonstrated a mean increase in circumference of the native aortic tissue of 0.67 cm (p value <0.01), whereas there was no significant change in homograft circumference.

Conclusions. After reconstruction of the aortic arch in HLHS, the diameter of the arch continues to increase throughout childhood, and this increase is due to growth of the native aortic tissue. (J Am Coll Cardiol 1998;32:1951–4)

©1998 by the American College of Cardiology

Hypoplastic left heart syndrome (HLHS) is marked by underdevelopment of the left-sided structures of the heart including the aortic arch. In the first stage of surgical palliation, the Norwood operation, the aortic arch is reconstructed by anastomosis of the native aorta to the main pulmonary artery and augmentation of the arch with pulmonary homograft (1). The goal of initial arch reconstruction is to create a large, unobstructed pathway for systemic circulation. Native aortic tissue is included in the neoaorta, although it accounts for only a small portion of the vessel’s circumference. This technique preserves the origins of the branch vessels and allows for potential growth of the neoaorta.

Little is known about the capacity for growth in the reconstructed aortic arch, yet an understanding of this capacity is important for several reasons. If there is little change in the caliber of the vessel after the Norwood operation, then there is potential for significant narrowing later in life. Conversely, significant growth of the neoaorta may have adverse effects on long-term outcome. Previous work has demonstrated that a large neoaorta can limit the growth of the left pulmonary artery (LPA) by trapping it against the left mainstem bronchus and causing LPA stenosis (2).

The purpose of this study is to determine whether the neoaorta grows after palliation for HLHS and whether this growth is due to expansion of the homograft or true growth of the native aortic tissue.

Methods

To investigate the growth characteristics of the neoaorta, we retrospectively examined the serial echocardiograms of intermediate-term survivors of the Norwood operation. The eligible patient population consisted of patients with HLHS who underwent a Norwood operation at The Children’s Hospital of Philadelphia. We identified 50 subjects in whom three good quality serial echocardiograms were available. The medical records for each patient were reviewed to obtain information about the year of surgery, surgeon and the incidence of coarctation and LPA stenosis. For each patient three echocardiographic studies were analyzed. The first study was obtained after the Norwood operation and before discharge from the hospital (mean age of 19.9 days, range 2 to 46 days). The second study was obtained beyond 1 year of age, most commonly at the time of the Fontan operation (mean age 2.5 years, range 1.0 to 7.2 years). The third study consisted of the most
Echocardiographic studies were performed using Hewlett-Packard Sonos 1500 (Andover, MA) Ultrasound Imaging System or Acuson 128XP/10c Computed Sonography (Mountainview, CA). A variety of transducer frequencies offering the best image resolution for age was used. Morphometric measurements were made in triplicate using off-line analysis with the Graphic Overlay System from Digisonics (Houston, TX). Measurements were obtained of the transverse arch diameter. Evaluating this portion of the aorta offers several advantages: 1) both native aortic tissue and pulmonary homograft are included in the transverse portion of the reconstructed arch; 2) the transverse arch has well-defined landmarks that can be visualized from the suprasternal notch; 3) the transverse arch is perpendicular to the ultrasound beam when viewed from the suprasternal notch, allowing for maximal image resolution and reproducibility, and 4) published values are available for the diameter of the transverse arch in the normal population (3).

We measured the diameter of the transverse arch in a long-axis suprasternal view between the origin of the innominate and the left carotid arteries. All measurements were obtained in diastole. Body surface area at the time of echocardiographic study was recorded.

We questioned how the caliber of the transverse neoaorta immediately after the Norwood operation relates to the diameter found later in life after the Fontan operation. For example, does a large neoaorta in infancy result in a large neoaorta in childhood? The size of the transverse arch in the neonatal period was compared to that measured at the most recent study. Because the follow-up studies were obtained at different ages, we employed a statistical model that takes into account both age and body surface area to test our hypothesis.

We investigated whether the growth of the neoaorta was due to true growth of the native aortic tissue or dilation of the pulmonary homograft. To do so, we evaluated autopsy specimens and echocardiograms of patients with HLHS (not included in the above group) who underwent the Norwood operation and expired after 1 year of age. Autopsy specimens and adequate echocardiographic studies were available from 10 patients. The echocardiograms of these patients obtained before (unoperated arch) and early after the Norwood operation were reviewed. The mean age of death of these subjects was 20.0 months (14 to 34 months). None of the subjects had significant coarctation of the aorta at autopsy. In these specimens, the transverse arch was incised in the long axis, and the circumference of the arch was measured between left innominate and left carotid arteries. In examining the specimens, it was possible to distinguish between native aorta and homograft, since there was a distinct difference in texture between the two tissues, and the suture lines were well demarcated. The relative contribution of the native aorta and homograft to the overall circumference of the transverse arch was measured. To assess the growth of the neoaorta in these patients the circumference of the transverse arch at the time of death was compared to the circumference of the transverse arch calculated from the echocardiogram obtained early after the Norwood operation (range, postoperative day 0 to 5). The circumference of the transverse arch after the Norwood operation was determined by the geometric equation: circumference = πd (where d is the diameter of the transverse arch in long-axis view). To assess the growth of the native aortic tissue alone, we measured its contribution to the transverse arch at the time of death and compared it to the echocardiographic measurement prior to the Norwood operation. Lastly, we compared the contribution of the homograft portion of the transverse arch at the time of death to that measured immediately after the Norwood operation. The postoperative measurement was calculated by subtracting the contribution of the native aortic arch to the neoaorta. At our institution the entire portion of the hypoplastic arch was retained in the construction of the neoaorta whenever possible. For the purposes of this study, we assumed that all of the native aortic tissue was included in the anastomosis.

Statistical analysis. To determine if the caliber of the neoaorta in infancy predicted the caliber later in life, we employed the repeated measures analysis of variance technique. Statistical analysis of differences between postoperative and postmortem measurements of arch circumference was performed with Student t test. Significance was determined at a p value <0.05. Statistical tests were performed with BMDP5V from BMDP Statistical Software Inc. (Los Angeles, CA).

Results

Patient characteristics. The 50 patients included in this study underwent the Norwood operation between April 1984 and December 1992. Two surgeons performed the Norwood operation at our institution, with one surgeon performing 45 of the 50 cases (90%). Review of medical records for those patients evaluated by serial echocardiography reveals that six patients underwent balloon dilation of aortic coarctation. The mean age at which this procedure was performed was 7.9 months. No patient had a residual gradient greater than 10 mm Hg after balloon dilation. None of the patients in our series underwent balloon dilation beyond 2 years of age, and none underwent surgical reconstruction of the neoaorta. Review of the angiography demonstrates that 12 of 50 patients (24%) had evidence of either LPA stenosis or LPA hypoplasia.

The incidence of late LPA stenosis could not be determined, since less than half the patients underwent cardiac catheterization after the Fontan operation.
Growth characteristics of the neoaorta: analysis of intermediate-term survivors. In each of the 50 intermediate-term survivors the transverse neoaortic arch increased in diameter with increasing body surface area. Plotting all measurements of transverse arch diameter against body surface area is demonstrated in Figure 1. The growth curve of the transverse arch in a normal pediatric population is provided for comparison.

When one examines the relationship of the neoaortic arch diameter in infancy to that in later childhood, one finds a statistically significant association. The initial arch measurement predicts the follow-up measurements (estimate 0.46, p value 0.001). These data suggest that the relative size of the neoaorta at the time of the Norwood operation impacts significantly on its ultimate size.

Contribution of components to growth of the arch: postmortem analysis. As one would expect from the serial follow-up data the caliber of the neoaorta increased significantly in the 10 subjects evaluated at autopsy. The circumference increased from 3.64 cm at the time of the Norwood operation to 4.46 cm at the time of death (Table 1). As well, the native aorta grew significantly from a mean of 1.19 cm prior to Norwood operation to 1.86 cm at the time of death. There was no significant change in the mean circumference of the homograft.

**Discussion**

The success of the Norwood operation as a palliative procedure depends in large part upon reconstruction of the aortic arch to provide adequate and unobstructed systemic blood flow. In the short term, a small caliber neoaorta can impede the function of the right ventricle and lead to hemodynamic instability. How the caliber of the neoaorta affects long-term outcome is less clear, since the growth of the neoaorta after the Norwood operation has not been described previously. In our study, we investigated the growth characteristics of the reconstructed arch after the Norwood operation for HLHS.

Previous publications have examined the growth of hypoplastic segments of the aorta in related lesions. Studies in patients with coarctation of the aorta and transverse arch hypoplasia have demonstrated that the hypoplastic segment grows after surgical repair of the discrete coarctation (4–6). Relief of obstruction at the site of the coarctation allows for increased flow through a hypoplastic transverse arch, which is thought to stimulate growth in vessel diameter. In these patients, the transverse arch growth after repair of coarctation is even more pronounced than in the normal population (6). The most rapid growth of the arch occurred within 6 months of repair. This would suggest that the potential for vessel growth is not affected adversely by diminished flow in utero. Growth of the hypoplastic aortic arch in subjects with valvar or subvalvar aortic stenosis has also been described. Turley evaluated patients with hypoplasia of the ascending and transverse aorta and either transposition of the great arteries, subaortic stenosis or interrupted aortic arch (7). He demonstrated that when increased flow is achieved through the proximal aorta with a pulmonary artery to descending aorta conduit and banding of the distal main pulmonary artery, the ascending and transverse aorta exhibit significant growth.

We have demonstrated that the neoaorta in HLHS also has the potential for significant growth. The rate of growth of the transverse arch after reconstruction appears to parallel that described in the normal population. Examination of the autopsy specimens suggests that this increase in diameter is due to growth of the native aortic tissue. Dilation of the homograft, conversely, does not play a significant role in the increased diameter of the neoaorta, at least in early childhood. The stimulus for growth of native aortic tissue is likely to be the increased flow of blood through the vessel. It has been proposed that in lesions such as HLHS, the ascending aorta and aortic isthmus become hypoplastic as a result of diminished flow through the left ventricular outflow tract in utero (8). Once the flow through the aortic arch is increased, as in the Norwood operation, the aortic tissue is stimulated to grow. The relationship between blood flow and vessel growth has been elaborated by the hemodynamic molding theory, whereby blood vessels grow around a column of blood (9,10). This

![Figure 1. Plot of transverse arch diameter versus body surface area (BSA) for 50 patients with hypoplastic left heart syndrome after Norwood operation at the 1st (circles), 2nd (triangles) and 3rd (diamonds) echocardiographic studies. The solid line represents the regression line for a normal population (Snider et al. [3]). The hatched lines are the tolerance limits weighted for body surface area for prediction of normal values for 80% of the future population with 50% confidence.](image-url)
theory has been validated in animal experiments where shear stress and stretch appear to be the two factors most responsible for inducing growth of large caliber arteries (11–13). Because large caliber vessels have the capacity to grow in response to increased flow, retaining the native aortic tissue in the reconstructed arch should allow for long-term growth.

There is also some evidence to suggest that the growth of vascular tissue is mediated by a variety of circulating growth factors that contribute to general somatic growth (11). Growth factors, such as insulin-like growth factor-1, have been shown to stimulate growth of cardiomyocytes and large vessels (14). It is interesting to note that in the patients with HLHS, the neoaorta continues to grow at a rate similar to the normal population, maintaining a larger caliber throughout childhood. If the quantity of blood flow through a vessel were the sole stimulus for growth, one would expect the caliber of the neoaorta to approach that of the normal population. However, this is not the case, suggesting somatic growth factors as well as mechanical forces play an important role in arch growth. This is further supported by our finding that initial arch diameter in the neoaorta relates to arch diameter later in life.

Our understanding of the growth potential of the reconstructed transverse arch has important clinical implications. Recently some centers have modified the Norwood operation, so that homograft is not employed in reconstruction of the aortic arch (15). One of the arguments supporting this approach has been the concern that the neoaorta will not grow. Our data suggest that the arch does grow after the Norwood operation. Residual coarctation of the aorta can develop as a result of failure to extend the homograft far enough around the arch or problems related to the suture line. However, significant narrowing of the neoaortic arch after the Norwood operation should not occur as a result of impaired growth of native aortic tissue.

Conversely, growth of the neoaorta can have some unfavorable effects on long-term outcome. Previous publications have described an asymmetry in the size of the pulmonary arteries, LPA smaller than right pulmonary artery, when a modified right Blalock–Taussig shunt is employed in the Norwood operation (2). In our population 24% of patients had angiographic evidence of LPA stenosis or hypoplasia. In many cases this is a result of compression of the PA between the insertion of the shunt and the LPA from the large neoaortic arch gusset. Such compression leads to diminished flow into the LPA, which in turn results in hypoplasia of that segment of the arterial tree. In Fontan physiology, where pulmonary blood flow is passive, the compression of the central pulmonary arteries by the neoaorta could also lead to diminished flow to the LPA. Since it is clear that the neoaorta continues to grow throughout childhood, the potential LPA stenosis might still exist even years after the completion of palliative surgery.

It is not possible to say with confidence what the fate will be of the reconstructed arch if children undergo the Norwood operation and survive to school age. The oldest child in the group was 13.6 years at the time of the most recent study. As well, the long-term durability of pulmonary homograft is not fully understood. Although experience suggests that the integrity of the homograft is quite good, the potential for aneurysmal changes does exist. Continued evaluation of the reconstructed arch in these patients as they reach adolescence and adulthood will be necessary to observe for potential complications and to characterize more completely the fate of the reconstructed aortic arch.

References