Aortic Arch Obstruction After the Norwood Procedure for Hypoplastic Left-Heart Syndrome

Is it Inevitable? Is it Preventable?*

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William I. Norwood, working at the Boston Children’s Hospital, developed and refined a surgical procedure that permitted babies born with hypoplastic left-heart syndrome (HLHS) to survive and enjoy life (1). The high mortality rates reported during the 1980s have gradually declined as various surgeons and medical centers have refined both the surgical techniques and the pre- and post-operative care for infants born with HLHS. In this issue of the Journal, Porras et al. (2) report on the treatment of aortic arch obstruction (COA) after the Norwood procedure (NP) (2). Their report specifically focuses on infants who underwent primary balloon dilation (BD) of COA after the NP. Infants requiring surgery as the primary treatment for COA after the NP were excluded from the analysis. The report is notable for multiple reasons. The study group was culled from the overall Boston Children’s Hospital NP experience from January 1993 to May 2009. During the time period studied, 556 patients underwent the NP at Children’s Hospital Boston. There were 462 patients alive at >30 days after surgery, and 133 required arch interventions (29%). Seventeen patients were excluded from the analysis, 13 presumably underwent primary surgical treatment, and 4 patients were excluded because primary BD was performed elsewhere or a stent was implanted. The impact of COA on early mortality (<1 month) was not examined in this analysis.

In the patient cohort analyzed, the indication for BD varied depending on the clinical findings and their consequences. A strong indication for BD was defined as a coarctation index <50% and/or a peak gradient ≥15 mm Hg in the presence of normal ventricular function or a gradient >10 mm Hg in the presence of “significant” ventricular dysfunction. Significant ventricular dysfunction was defined as moderate or worse ventricular dysfunction on pre–cardiac catheterization echocardiography or evidence of hemodynamic compromise (i.e., ventricular end-diastolic pressure ≥14 mm Hg and/or calculated cardiac index <2.6 l/min/m²). These indications for intervention are similar to the criteria used for intervention for COA after the NP at Rady Children’s Hospital San Diego.

This report is a very important contribution not only because it emanates from the medical center where the NP was developed and refined but also because the investigators have provided demographic information that permits an estimate of the overall incidence of important COA after the NP. Recently, the Pediatric Heart Network (PHN), a consortium of 15 North American medical centers focused on the treatment of congenital heart disease, reported the results of a randomized clinical trial designed to compare shunt types after the NP for single-ventricle lesions (3). The Boston Children’s Hospital is a member of the PHN. In the PHN trial, 12-month survival was 73.7% in the right ventricle–to–pulmonary artery (RVP A) shunt group and 63.6% in the modified Blalock-Taussig (MBT) shunt group. Of note, 4.7% of the RVPA patients required revision of the neoaorta during the initial Norwood hospitalization, while 2.5% of the MBT patients required arch interventions during the first hospitalization. In the interval between Norwood discharge and stage II discharge, 21.5% of the RVPA shunt patients required arch interventions, while 18.7% of the MBT shunt patients required interventions. Finally, between stage II discharge and 12 months of age, an additional 6.8% of the RVPA shunt patients required interventions, while 4% of the MBT shunt patients required interventions. These findings suggest that early and late arch obstruction occurs in a substantial number of patients undergoing the NP at the leading institutions focused on the treatment of patients with HLHS in North America.

What lessons can be learned from the study by Porras et al. (2)? The investigators looked for potential causal factors for COA requiring intervention. They found that COA after the NP could be categorized as proximal or distal. The most common site of COA was distal to the left subclavian artery (88% of patients). Fourteen patients had proximal COA. Ten of 14 had obstructions in the transverse arch between the origins of the innominate and left subclavian arteries. Four patients had COA proximal to the origin of the brachiocephalic vessels. Proximal COA was significantly associated with shorter freedom from reintervention, reoperation, and transplantation-free survival. Patients with...
proximal COA underwent initial BD during unplanned catheterization more often than patients with distal COA. Symptoms due to arch obstruction also predicted a poor outcome. Overall, BD was acutely successful in most patients. However, reintervention, either catheter based or surgical, after primary BD for COA after NP was necessary in many patients.

Potential surgical and technical factors associated with COA after NP were analyzed. There was no association between individual surgeons and patient outcomes. Some investigators (4,5) have suggested that coarctectomy at the time of arch reconstruction during the NP portends an improved late outcome. In the study by Porras et al. (2), coarctectomy at the time of the NP did not influence the outcome of BD for late COA. In a different report from Boston Children’s Hospital, Bautista-Hernandez et al. (6) described outcomes in a smaller cohort of patients operated between January 2000 and June 2005. In their report, 210 patients who underwent the NP were analyzed. Operative mortality was 10%. The incidence of COA requiring intervention (BD, surgical arch augmentation, or both) was reported as 24%. Pre-operative anatomic aortic coarctation was consistently linked to late neoaortic arch obstructions. In the study by Bautista-Hernandez et al. (6), patients having aortic arch repair by means of direct connection (i.e., coarctectomy) with or without autologous pericardium patching at the NP were less likely to have late COA. It is very interesting that in a somewhat different analysis of a group of patients that included those reported by Bautista-Hernandez et al. (6), Porras et al. (2) conclude that coarctectomy at the time of the NP is not protective in the subset of patients who develop late COA.

What is the take-home message of this report? First, data from this single-center report and the PHN pooled data indicate that the problem of late COA after the NP has not been solved. Porras et al. (2) were not able to identify anatomic factors that correlated with their outcomes, including examination of the shape of the transverse arch on the post-operative angiograms to determine if unusual arch anatomy predicted late problems. There were not enough patients with aberrant right subclavian arteries to permit separate analysis of that subgroup. Porras et al. do not comment on other variations in the branching of aortic arch vessels that may be associated with a small transverse aortic arch or the absence of an aortic isthmus.

It is likely that late distal COA will be inevitable in a small subset of patients undergoing NP for several reasons. Unusual anatomy of the distal transverse arch and/or the isthmus of the aorta, particularly in the presence of an aberrant subclavian artery, may preclude effective coarctectomy. In that situation, there may be a very narrow strip of native tissue, and an onlay patch must be used. In addition, it is difficult, if not impossible, to identify the entire geography of the ductal tissue in the operating room. Surgeons performing the NP normally use optical assistance and visual cues to determine where ductal tissue ends and normal aorta begins. Residual ductal tissue inadvertently incorporated into the repair can result in progressive involution and narrowing of the distal portion of the arch reconstruction. If this anatomic substrate is present, a subset of patients undergoing the NP will develop distal COA simply because it is impossible to surmount anatomic obstacles presented by the arch configuration or the location of ductal tissue. Late distal arch obstruction may respond favorably to BD because the interventionalist is dilating constricted ductal tissue rather than a circumferential surgical scar. In contrast, proximal arch obstruction is probably always related to technical factors. Proximal COA caused by an inadequate or kinked patch is less likely to be amenable to BD. Can proximal arch obstruction and/or transverse arch obstruction be prevented? Intraoperative transesophageal echocardiography may not yield satisfactory views of the ascending aorta and transverse arch. If right radial and femoral or umbilical artery pressures are routinely monitored during the NP, a gradient noted at the completion of operation would identify transverse arch obstruction and permit immediate revision. Passing the right atrial catheter across the tricuspid valve and comparing right ventricular systolic pressure with femoral or umbilical artery pressure will permit an evaluation of all the potential areas of stenosis after the repair. Of course, on-the-table angiography performed in a hybrid operating room would also identify areas of narrowing likely to become early or late COA sites. It is also likely that identification of proximal or distal COA immediately after NP might affect the reported early mortality rate.

In summary, the report by Porras et al. (2) is an important contribution because it defines an ongoing problem in the surgical treatment of HLHS. The PHN report confirms that the problem of early and late COA after the NP for HLHS still occurs at the leading centers treating congenital heart disease in North America. Fortunately, many patients can be helped by BD or surgery. Proximal COA may be identified and corrected at the time of NP if the post-NP intraoperative analysis is aggressive and thorough. Although the prevention of distal COA remains a challenge, it is reassuring to note that in a report by Ballweg et al. (7), reintervention for late COA did not affect short to midterm outcomes during the staged approach to treatment of HLHS.

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