Pre–stage II mortality after the Norwood operation: Addressing the next challenge

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Achieving consistently good operative survival after the Norwood operation can be considered an "acid test" of sorts for a pediatric cardiovascular care program. All the provider components of the program are put to the test. Before the operation, these patients must be managed in the intensive care unit with careful management of the balance between systemic and pulmonary blood flow. Patients in whom ductal closure has precipitated near or bona fide circulatory collapse require skillful resuscitation. During the operation, skill and precision are required of surgeon, anesthesiologist, and perfusionist. Volumes have been written about the postoperative challenges presented in the intensive care unit by these neonatal patients, who have been subjected to cardiopulmonary bypass, myocardial ischemia, and, in some cases, whole-body ischemia. Importantly, they emerge from this operation *without* a repair. This is a single-ventricle circulation, dependent on a systemic-pulmonary shunt for pulmonary blood flow. Maintaining an appropriate balance between systemic perfusion and pulmonary blood flow can again prove difficult. Obtaining adequate cardiac output from the postischemic single ventricle with a coronary circulation that is subjected to continuous diastolic runoff into the lungs through the modified Blalock-Taussig shunt may also be a challenge. It is therefore not surprising that an institutional surgical mortality for the Norwood operation of 10% to 20%, an order of magnitude higher than that seen for many complex neonatal biventricular repairs, can still be considered a fine achievement.

The number of institutions reaching this level of excellence is steadily increasing. In this issue, Ghanayem and colleagues[1] present additional data from the ongoing experience with the Norwood operation at Children's Hospital of Wisconsin in Milwaukee. Dr James Tweddell and his associates there have indeed achieved excellent surgical results with the Norwood operation. Here they report 93% hospital survival in 87 consecutive Norwood operations since 1996. These results are admirable. This surgical group and others, however, have noted that a number of patients who survive hospitalization after the Norwood procedure, still in possession of a potentially unstable shunt-dependent single-ventricle anatomic arrangement, die before returning for the second stage, bidirectional cavopulmonary anastomosis. This pre–stage II mortality has been reported to approach 10% to 15% of survivors of the initial Norwood operation.[2] It is important to note that although some patients dying before stage II are noted at autopsy to have residual defects, such as coarctation or shunt stenosis, many have no identifiable problem with their Norwood anatomy.

In their article "Home Surveillance Program Prevents Interstage Mortality After the Norwood Procedure," Ghanayem and colleagues[1] report the results of an aggressive programmatic response to this pre–stage II mortality. *Pre–stage II mortality* is used here rather than *interstage mortality*, because it is during that time frame that these patients are at risk for out-of-hospital death. Out-of-hospital death is rare after second- and third-stage palliation. The Milwaukee investigators discharged all survivors of the Norwood operation from the hospital with infant scales and pulse oximeters. They hypothesized that daily home surveillance of patient weight for identification of dehydration or growth failure and of arterial oxygen saturation for the identification of potentially dangerous levels of desaturation might decrease pre–stage II mortality among their patients. Parents of these survivors were

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advised to seek medical advice in case of room air oximetric saturation less than 70%, weight loss greater than 30 g, or failure to gain 20 g during the course of 3 days. Twenty-four patients treated with this home surveillance program since 2000 were compared with a historical cohort of 63 patients treated from 1996 to 2000. Ghanayem and colleagues[1] report a 16% pre–stage II mortality in the historical cohort versus no pre–stage II deaths in the group subjected to home surveillance. Patients who met the warning sign criteria underwent the second-stage bidirectional cavopulmonary anastomosis at a younger mean age (3.7 months) than did those who never met those criteria, who underwent bidirectional cavopulmonary anastomosis at a mean age of 5.2 months. Ghanayem and colleagues[1] concluded that the earlier progression of these "at-risk" survivors probably contributed to their decreased pre–stage II attrition.

Several important observations can be made when looking at these data. First, these clinical results are truly excellent. They were obtained with what will soon be known as a "classic" Norwood operative strategy. A minirevolution is currently taking place with regard to the first-stage palliation of patients who would have previously undergone a Norwood operation with a modified Blalock-Taussig shunt. Largely because of the weaknesses described here, many surgeons are now providing pulmonary blood flow at the time of stage I palliation with a single ventricle–pulmonary artery conduit. This procedure, pioneered by Sano and colleagues,[3] appears to improve the early postoperative hemodynamics of these patients. In particular, the diastolic blood pressure of these patients is considerably higher than that seen in patients with a modified Blalock-Taussig shunt. Sano, and now others, are reporting favorable early outcomes after this type of first-stage palliation. Dr Sano reports that this modification has dramatically improved survival after the Norwood operation at his institution.

When one looks at the Norwood results from Ghanayem and colleagues,[1] however, they leave little room for improvement. At this time it might not be prudent for their program to abandon the more classic Norwood operation for the Sano modification, which carries with it the soon-to-be-defined consequences of a systemic ventriculotomy in these single-ventricle patients. Proponents of the Sano modification must, at some point in the future, compare their results with classic Norwood results, such as those presented here.

A second observation involves the fact that, as the authors acknowledge, these data are retrospective, and the control group was historical. Only a significantly larger and longer home-monitored "treatment group" experience or a prospective randomized trial could more confidently assert that home monitoring will essentially eliminate pre–stage II mortality. That said, however, one cannot deny that, for whatever reason, pre–stage II death has become exceedingly rare during the past several years in this program. This inability to identify a single causative factor for such an improvement in outcome in a complex system such as this was first described by Elton Mayo of the Harvard Business School in his landmark studies at the Western Electric Hawthorne Works in Cicero, Illinois, between 1927 and 1932.[4] Mayo and his colleagues, in an unblinded fashion, were examining the effects of various environmental conditions on worker productivity. To their amazement, these early industrial psychologists discovered that the production of the workers improved regardless of the specific experimental manipulation of workplace variables. The experimental group of workers had been segregated from the general workforce population, and the changes in the environmental variables were explained to them in discussions before the period of data collection. The investigators noted that the workers developed an increased sense of responsibility, and the assembly line team became a tighter social entity. This beneficial "distortion" of results caused by the special attention human experimental subjects receive from researchers has come to be known as the "Hawthorne effect" and forms one of the pillars of human relations management and industrial psychology.

It is not a stretch to see the home-monitoring parents in Dr Ghanayem's study as Mayo saw the Western Electric factory workers. The overt enlistment of the family in the pre–stage II home monitoring care of these patients undoubtedly increased the sophistication level of postoperative and predischarge teaching. It also likely made the family feel more empowered to contribute positively to the care of their child. Rather than speculate on what the causative reason or reasons for this decrease in pre–stage II mortality, one should give Mayo his due and accept at least some part of this as a result of the Hawthorne effect. Finally, one must direct praise to these clinicians at the Children's Hospital of Wisconsin. They have shown us that, with what now appears to be appropriately increased programmatic attention to this problem, pre–stage II mortality can be dramatically reduced.

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