Association between Technical Performance Scores and neurodevelopmental outcomes after congenital cardiac surgery

Meena Nathan, MD,[a] Anjali Sadhwani, PhD,[b] Kimberlee Gauvreau, ScD,[c] Michael Agus, MD,[d] Janice Ware, PhD,[b] Jane W. Newburger, MD, MPH,[b,c] and Frank Pigula, MD[a,b]

Objectives: Technical Performance Score (TPS) has been shown to have a strong association with early and late outcomes after congenital cardiac surgery, with greater morbidity and reintervention in children with major residual lesions (TPS class 3). We sought to explore the effect of TPS on the neurodevelopmental outcomes.

Methods: All infants undergoing cardiac surgery, excluding those with trisomy 21, were offered neurodevelopmental testing at 1 year of age using the Bayley Scales of Infant Development, 3rd edition. TPSs from the discharge echocardiograms were graded as class 1 (optimal), class 2 (minor residual), or class 3 (major residual). Multivariate regression analysis was performed using patient characteristics and preoperative variables.

Results: Neurodevelopmental testing was performed in 140 patients at a median age of 16 months. Of these, 28 (20%) had single ventricle palliation; 39 (28%) were in Risk Adjustment for Congenital Heart Surgery category 4 to 6. Significant differences between the groups were found in the cognitive ($P = .01$) and motor ($P = .05$) domains, with subjects in TPS class 3 having significantly lower cognitive and motor composite scores. The scores did not vary significantly according to single ventricle versus biventricular repair or Risk Adjustment for Congenital Heart Surgery categorization. In multivariate modeling, class 3 TPS remained significantly associated with a lower Bayley cognitive score ($P = .02$), with a trend toward a lower Bayley motor score ($P = .08$).

Conclusions: We found that TPS is an independent predictor of neurodevelopmental outcomes after infant heart surgery. Future research should explore whether a structured program of intraoperative recognition and intervention on residual lesions can improve the TPS and neurodevelopmental outcomes. (J Thorac Cardiovasc Surg 2014;148:232-7)

CHD

Supplemental material is available online.

Congenital heart disease is the most common birth defect, occurring in 9/1000 live births, one third of whom require intervention in infancy.[1,2] Although the survival rates have significantly increased, numerous studies have documented the neurodevelopmental (ND) deficits experienced by children with congenital heart defects across different ages.[1-8]

It is well known that the outcomes after surgery for complex congenital cardiac defects are dependent on multiple factors, including disease complexity, preoperative severity of illness and physiologic status, intraoperative conduct of the operation, and postoperative course, including length of hospital stay, care in the intensive care unit, and events requiring intervention before discharge.[9,10] Among these multiple factors, the technical adequacy of the repair might be the single most important factor determining outcomes. In previous work at our institution,[11,12] we have developed and validated a scoring system termed the "Technical Performance Score" (TPS) that scores the adequacy of the repair using specific echocardiographic and clinical criteria at discharge. We have shown that class 3 (inadequate TPS, major residual defect often requiring predischarge surgical or catheter-based reintervention on the treated anatomic area) was strongly associated, not only with early mortality and complications, but also with significantly increased resource use, as measured by ventilation time, hospital length of stay, and hospital charges after adjusting for patient-specific factors such as age, presence of chromosomal or other nonchromosomal abnormalities, prematurity, and disease complexity.[13-15] We were also able to show that this significant association between class 3 (inadequate) TPS persisted during mid-term follow-up for mortality and the need for late reintervention.[16,17]

We hypothesized that a strong association would exist between class 3 (inadequate) TPS and poorer ND outcomes (NDOs). The present study investigated the relationship

From the Department of Cardiac Surgery,[a] Cardiac Neurodevelopmental Program,[b] Department of Cardiology,[c] and Medicine Critical Care Program,[d] Boston Children's Hospital and Harvard Medical School, Boston, Mass.

Disclosures: Authors have nothing to disclose with regard to commercial support.
Presented as a moderated poster presentation at the American College of Cardiology 62nd Annual Scientific Session, San Francisco, California, March 9-11, 2013.
Received for publication May 3, 2013; revisions received July 29, 2013; accepted for publication Aug 11, 2013; available ahead of print Sept 30, 2013.

Address for reprints: Meena Nathan, MD, Department of Cardiac Surgery, Boston Children's Hospital and Harvard Medical School, 300 Longwood Ave, Bader 273, Boston, MA 02215 (E-mail: meena.nathan@cardio.chboston.org).

0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2013.08.032

Abbreviations and Acronyms

BSID	= Bayley Scales of Infant Development, 3rd edition
ECMO	= extracorporeal membrane oxygenation
ICU	= intensive care unit
ND	= neurodevelopmental
NDO	= neurodevelopmental outcomes
RACHS-1	= Risk Adjustment for Congenital Heart Surgery
TPS	= Technical Performance Score

between the TPS and NDOs in a group of infants who had undergone a wide variety of congenital cardiac operations in the first year of life and returned for ND follow-up from 14 months to 3 years of age.

METHODS

A retrospective chart review of all infants <1 year old, who had undergone surgery from August 2008 to September 2011 and who had undergone ND testing at our center, was performed with institutional review board approval. Patients with trisomy 21 were excluded because of the known developmental delays associated with that population.

Technical Performance Score

Every patient had a TPS determined using discharge echocardiographic findings and clinical status at discharge from the index operation. The procedures were subdivided into components. Each component was scored as class 1 (optimal, no residual defect), class 2 (adequate, minor residual defect), or class 3 (inadequate or major residual defect). Any unplanned predischarge reintervention on the anatomic area repaired at the index surgery for a major residual defect resulted in a class 3 TPS. The final score for the procedure was determined by the summation of the subprocedure scores. The final score was deemed class 1 (optimal) if all subprocedure scores were optimal, class 2 (adequate) if ≥1 subprocedure scores were adequate, and class 3 (inadequate) if ≥1 subprocedure scores were inadequate.

ND Testing

ND testing was conducted using the Bayley Scales of Infant Development, Third Edition (BSID), which was administered by a psychologist. The BSID is a standardized measure that evaluates the current development of infants and children ≤3.5 years old. Composite scores (mean ± standard deviation, normal range 100 ± 15) were reported for the cognitive, language, and motor domains. In addition, the parents completed the Bayley Social-Emotional and Adaptive Behavior Questionnaire, a standardized questionnaire assessing the social and emotional development and adaptive functioning of children. Composite scores (mean ± standard deviation, normal range 100 ± 15) were reported for the overall social and emotional and adaptive functioning.

Risk Adjustment for Congenital Heart Surgery Risk Categories

The Risk Adjustment for Congenital Heart Surgery (RACHS-1)[18,19] method was developed to adjust for baseline case mix differences in risk when comparing in-hospital mortality among groups of patients aged <18 years undergoing congenital heart surgery. A nationally representative 11-member panel of pediatric cardiologists and cardiac surgeons used clinical judgment to place surgical procedures into 6 groups with a similar risk of in-hospital mortality; these risk categories were then refined using empirical data from the Pediatric Cardiac Care Consortium and 3 statewide hospital discharge databases. Category 1 has the lowest risk of death and category 6, the highest. Patients with combinations of cardiac surgical procedures (eg, repair of coarctation of the aorta and ventricular septal defect closure) should be placed in the risk category corresponding to the single highest risk procedure. The RACHS-1 risk categories have been widely validated for the outcome of in-hospital mortality and have also been used with other outcomes, including complications and total hospital charges.

Statistical Analysis

The patient and procedural characteristics are summarized using the median and range for continuous variables and the frequency and percentage for categorical variables. The mean ND scores were compared across TPS categories using 1-way analysis of variance. Because ND testing is voluntary in our center, we performed a sensitivity analysis to compare patients who had undergone ND testing with those who had not, to confirm the presence or absence of a selection bias in our study cohort. Relationships between the pre- and postoperative factors and TPS category were explored using the Kruskal-Wallis test for continuous variables and Fisher's exact test for categorical variables. Univariate associations between the BSID cognitive and motor composite scores and the pre- and postoperative factors were assessed using the unpaired *t* test or Spearman rank correlation coefficient. Only preoperative patient factors were considered for inclusion in the multivariate linear regression models. Previous work has demonstrated collinearity between the TPS category and intraoperative factors, such as the cardiopulmonary bypass time and aortic crossclamp time. Similarly, TPS is strongly associated with postoperative variables such as the need for extracorporeal membrane oxygenation (ECMO), ventilator duration, and intensive care unit (ICU) length of stay. We, however, did include the circulatory arrest time in our multivariate modeling, because previous large ND studies have shown a relationship between circulatory arrest and NDO. Preoperative factors significant at the $P = .20$ level were considered for inclusion, and the models were fitted using forward selection. Only factors significant at the $P = .10$ level were retained in the final model. Our primary goal was to assess the association between the TPS and the NDOs. Other covariates were considered to be potential confounders in the present analysis. Because covariates that have only a borderline significant relationship with the outcome can still be confounders, we chose a less stringent significance level of $P = .1$ for inclusion. The TPS category was then added to the model to determine whether it remained significantly associated with the NDOs after risk adjustment. Partial coefficients of determination were calculated for each variable in the final model. All analyses were performed using STATA, version 12 (StataCorp LP, College Station, Tex) for statistical analysis.

RESULTS

ND testing was performed in 140 infants (excluding those with trisomy 21) at a median age of 16 months. Of the 140 patients, 28 (20%) had single ventricle anatomy, 11 (8%) were diagnosed with known genetic conditions, 29 (21%) had additional noncardiac congenital anomalies, and 122 (87%) underwent surgery using cardiopulmonary bypass. Seven patients (5%) were premature (1 at 30 weeks and 2 each at 33, 35, and 36 weeks of gestation). The patient and procedural characteristics are listed in Table 1. All patients who underwent ND testing could be assigned a RACHS-1 category.

The mean BSID composite scores for the entire cohort were as follows: cognitive, 100 ± 17 (range, 43-145);

CHD

TABLE 1. Patient and procedural characteristics (n = 140)

Characteristic	Value
Male gender	77 (55)
Gestational age (wk)	39 (30-42)
Age at surgery (d)	34 (1-333)
Neonates	68 (49)
Weight (kg)	3.7 (1.4-9.5)
Noncardiac anomalies	29 (21)
Chromosomal anomalies	11 (8)
Single ventricle	28 (20)
Surgery with cardiopulmonary bypass	122 (87)
RACHS-1 risk category	
1	4 (3)
2	55 (39)
3	42 (30)
4	27 (19)
6	12 (9)
TPS	
Class 1 (optimal)	83 (59)
Class 2 (adequate)	44 (31)
Class 3 (inadequate)	13 (9)

Data presented as n (%) for categorical variables and as the median (range) for continuous variables. *RACHS-1*, Risk Adjustment for Congenital Heart Surgery; *TPS*, Technical Performance Score.

language, 94 ± 17 (range, 47-129); motor, 89 ± 17 (range, 46-130); social, 101 ± 21 (range, 55-145); and adaptive behavior, 91 ± 17 (range, 47-142). The distributions of the BSID cognitive and motor composite scores are presented in Figure 1.

The BSID composite scores across the TPS classes are listed in Table 2. Significant differences between the groups were found in the cognitive ($P = .01$) and motor ($P = .05$) domains, with subjects in TPS class 3 having significantly lower cognitive and motor composite scores than subjects in classes 1 and 2. Examining the relationship between TPS and the other pre- and postoperative medical variables, we found that class 3 TPS was associated with a higher RACHS-1 risk category. Similarly, patients with TPS class 3 had a greater need for ECMO, a prolonged ventilator duration, and longer ICU and hospital lengths of stay. These findings are listed in Table E1.

Univariate analyses were conducted to explore the relationships between potential predictor variables and the BSID cognitive and motor composite scores. The BSID cognitive composite score showed a significant association with chromosomal anomalies, the need for postoperative ECMO, feeding status at discharge, and postoperative intubation times (Table E2). The BSID motor composite score showed a significant association with chromosomal anomalies, repair type (ie, single ventricle palliation vs biventricular circulation), the need for postoperative ECMO, feeding status at discharge, and postoperative intubation time, and lengths of hospital and ICU stay (Table E3).

After adjusting for the presence of chromosomal anomalies, nonchromosomal anomalies, and RACHS-1 category 6, class 3 TPS remained significantly associated with a lower BSID cognitive composite score ($P = .02$). Anomalies and RACHS-1 risk category 6 together explained 12.5% of the variability in the cognitive composite score and TPS 3.9% (Table 3). Similarly, after adjusting for the presence of chromosomal anomalies, nonchromosomal anomalies, single ventricle palliation, and circulatory arrest time, class 3 TPS had only a borderline relationship ($P = .08$) with a lower BSID motor composite score (Table 4). The patient and operative characteristics together accounted for 13.1% of the variability in the motor composite score, with TPS adding another 2.0%.

All analyses were repeated, excluding the patients with known chromosomal anomalies. On multivariate analysis, class 3 TPS remained significantly associated with a lower BSID cognitive composite score ($P = .03$). It was not significantly associated with the BSID motor composite score (Tables E4 and E5).

DISCUSSION

We have previously shown that better technical adequacy of repair of congenital heart lesions, as measured by the TPS, has a positive effect, not only on early outcomes such as mortality, adverse events, and ICU and hospital

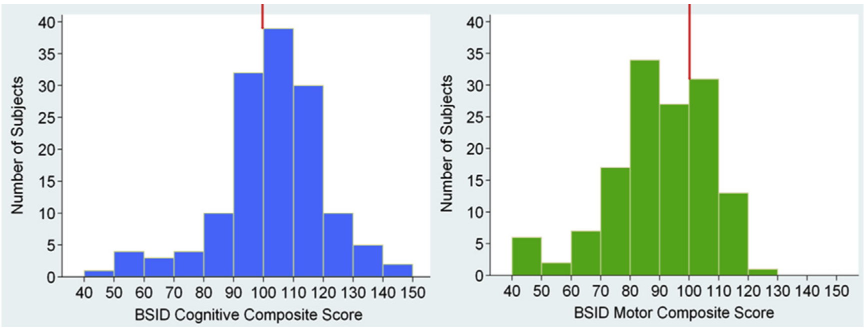

FIGURE 1. Distribution of Bayley Scale of Infant Development, 3rd edition (*BSID*), cognitive and motor composite score in the study subjects. The *red line* represents the mean value of the cognitive composite score in normal infants.

TABLE 2. Neurodevelopmental outcomes across Technical Performance Score classes

BSID III composite score	Class 1 (optimal; n = 83)	Class 2 (adequate; n = 44)	Class 3 (inadequate; n = 13)	*P* value
Cognitive	103 ± 17	99 ± 16	88 ± 22	.01
Language	95 ± 17	94 ± 17	85 ± 15	.16
Motor	91 ± 15	90 ± 17	79 ± 21	.05
Social	100 ± 22	101 ± 20	106 ± 20	.66
Adaptive behavior	89 ± 17	93 ± 18	93 ± 17	.47

Data presented as mean ± standard deviation. *P* values compare all 3 TPS classes. *BSID,* Bayley Scale of Infant Development; *TPS,* Technical Performance Score.

length of stay, but also on midterm mortality and the need for reinterventions.[13,14,16,17] Optimal TPS was not only associated with lower mortality and adverse events but was also able to compensate for any preoperative adverse physiologic effect.[13] However, the effect of TPS on neurodevelopment has not been previously studied.

We found that TPS class 3, indicating a technically inadequate operation, was an independent risk factor for a lower BSID cognitive composite score ($P = .02$) and tended to be associated with a lower BSID motor composite score ($P = .08$) in multivariate models adjusting for relevant patient factors and preoperative variables. These findings have extended the available data on the effects of TPS, showing that the technical adequacy with which an operation has been completed has an effect on the NDOs after congenital heart surgery. Children with significant residual lesions after repair should be considered at increased risk of developmental delay.

Our study has shown that the TPS accounts for 2% to 4% of the 15% variability in NDOs contributed by the covariates tested. The partial R^2 values relating the baseline patient characteristics to the NDOs have, in general, tended to be low.[20] However, the increase in the coefficient of determination resulting from the addition of TPS to the baseline patient factors was quite large in relative terms (a relative increase of 35% in the R^2 for the BSID cognitive composite score and a relative increase of 15% for the BSID motor composite score). More importantly, the magnitude of the effect was large. Even after controlling for confounding variables, patients with an inadequate TPS performed an average of 12.5 points lower on the BSID cognitive composite score than patients with an optimal TPS and an average of 8.5 points lower on the BSID motor composite score.

ND impairment, including attention deficit and hyperactivity disorder and executive dysfunction, are relatively common disabilities in children who have undergone surgery for congenital cardiac defects. Considerable data have documented the association of adverse NDOs with patient factors and preoperative variables, intraoperative management, and the postoperative course.[20-25] Some risk factors (eg, the presence of a genetic syndrome) are not modifiable. However, measures of greater global morbidity after cardiac surgery, such as a longer length of hospital stay or more adverse events before hospital discharge, increase the risk of ND delay.[23] To the extent that a more complicated postoperative course can be triggered by the technical inadequacy of surgery, an inadequate technical performance might be a modifiable risk factor for NDOs.

In our analysis, we chose to adjust only for those factors that could be measured up to the time when the TPS could be determined (ie, the preoperative and intraoperative factors). We chose not to include postoperative events such as periods of low cardiac output, cardiac arrest, and ECMO as covariates in the model, because these factors had a very strong collinearity with the TPS. Because the present study was not designed to distinguish between the effects of TPS and the immediate postoperative outcomes, we chose to use the TPS as a surrogate for these postoperative events. In our present cohort of patients, the representation of the lower risk RACHS-1 risk categories (categories 1-3) was greater, with the high-risk RACHS-1 categories representing about 28% of the total population. We have

CHD

TABLE 3. Multivariate analysis of Bayley Scale of Infant Development cognitive composite score adjusting for preoperative factors (total $R^2 = 14.6\%$)

	Preoperative and operative risk factors*		Preoperative and operative risk factors plus TPS		
Variable	Regression coefficient ± SE	*P* value	Regression coefficient ± SE	*P* value	Partial R^2 (%)
Chromosomal anomalies	−15.1 ± 5.3	.005	−14.4 ± 5.2	.007	5.4
Nonchromosomal anomalies	−10.4 ± 4.2	.02	−10.2 ± 4.2	.02	4.2
RACHS-1 category 6	−11.4 ± 5.0	.03	−7.7 ± 5.3	.15	1.5
TPS class (vs optimal)					
Adequate (class 2)	—		−2.4 ± 3.1	.45	
Inadequate (class 3)	—		−12.5 ± 5.1	.02	3.9

TPS, Technical Performance Score; *SE,* standard error; *RACHS-1,* Risk Adjustment for Congenital Cardiac Surgery. *Significant at $P = .10$ level; in the model on the right (with TPS), we see that an inadequate TPS was significantly associated with a lower score after adjusting for chromosomal and nonchromosomal anomalies and RACHS-1 category.

TABLE 4. Multivariate analysis of Bayley Scale of Infant Development motor composite score adjusting for preoperative factors (total R^2 = 15.3%)

Variable	Preoperative and operative risk factors*		Preoperative and operative risk factors plus TPS		
	Regression coefficient ± SE	P value	Regression coefficient ± SE	P value	Partial R^2 (%)
Chromosomal anomalies	−16.1 ± 5.1	.002	−15.6 ± 5.1	.003	6.7
Nonchromosomal anomalies	−8.0 ± 4.0	.05	−8.0 ± 4.1	.05	2.9
Single ventricle	−6.9 ± 3.4	.05	−5.9 ± 3.6	.11	2.0
Circulatory arrest	−7.2 ± 3.4	.04	−6.2 ± 3.4	.07	1.5
TPS class (vs optimal)					
Adequate (class 2)	—		−1.3 ± 3.1	.68	2.0
Inadequate (class 3)	—		−8.5 ± 4.9	.08	

TPS, Technical Performance Score; *SE,* standard error. *Significant at $P = .10$ level; in the model on the right (with TPS), we see that an inadequate TPS was only borderline associated with a lower score after adjusting for chromosomal and nonchromosomal anomalies, single ventricle palliation, and the use of circulatory arrest.

continued to enroll patients in our ND program, and in our next analysis, we would hope to have accrued a greater number of patients with higher complexity, which would allow us to establish associations between the TPS and NDOs among higher and lower complexity patients separately. We are also in the process of collecting data on the individual procedure groups to further refine this association.

Our study should be viewed in the light of certain limitations. This was a retrospective study. Not all the patients at our center who underwent cardiac surgery at <1 year of age returned for ND testing; the families whose residence was closer to our center were more likely to return. In addition, the families with children with more developmental delays might have been more motivated to return for testing. We performed a sensitivity analysis comparing those who had undergone ND testing (n = 140) with those who had not in 2011(n = 257; Table E6). That analysis demonstrated that no significant baseline difference were present between those who did and did not undergo ND testing, except for a greater rate of nonchromosomal anomalies in those who had undergone testing. In particular, the patients did not differ in age at surgery or surgical complexity. This demonstrated that a selection bias had not influenced the results of our analyses on the association of developmental delay with technical performance. Additional reinterventions after discharge but before ND testing might have had an influence on the final NDOs. Although we have shown in previous work that an inadequate TPS has an association with need for postdischarge reintervention, we did not include this factor in our analysis because of the small number of patients in the current analysis. Once data from a larger cohort of patients have been accrued, we plan to expand our analysis to include the postoperative and postdischarge factors that might contribute to the NDO.

ND testing at a young age (mean, 16 months) has limited predictive validity for later performance in normal children.[24] Moreover, the results from a recent study have suggested that the BSID will underestimate developmental delays.[8] Therefore, longitudinal follow-up data are needed to assess the effect of TPS on later NDOs. Finally, our data were gathered from a single center, potentially limiting the generalizability.

In conclusion, our data suggest that worse technical performance of infant heart surgery exerts an adverse effect on the NDOs after congenital heart surgery. In previous work, we have shown that intraoperative recognition and correction of residual lesions resulted in superior medical outcomes compared with those who required reintervention before discharge.[14] Future, prospective, multicenter research should explore whether a structured program of intraoperative recognition and intervention on residual lesions can improve the TPS and NDOs.

The authors would like to acknowledge Donna Donati and Jacqueline O'Brien for their contributions to the present study.

References

1. Karsdorp PA, Everaerd W, Kindt M, Mulder B. Psychological and cognitive functioning in children and adolescents with congenital heart disease: a meta-analysis. *J Pediatr Psychology*. 2007;32:527-41.
2. Marino BS, Lipkin PH, Newburger JW, Peacock G, Gerdes MJ, Gaynor W, et al. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. *Circulation*. 2012;126:1143-72.
3. Limperopoulos C, Majnemer A, Shevell ML, Roblicek C, Rosenblatt B, Tchervenkov C, et al. Predictors of developmental disabilities after open heart surgery in young children with congenital heart disease. *J Pediatr*. 2002;141:51-8.
4. Soto CB, Olude O, Hoffmann RG, Bear L, Chin A, Dasgupta M, et al. Implementation of a routine developmental follow-up program for children with congenital heart disease: early results. *Congenit Heart Dis*. 2011;6:451-60.
5. Fuller S, Nord AS, Gerdes M, Wernovsky G, Jarvik GP, Bernbaum J, et al. Predictors of impaired neurodevelopmental outcomes at one year of age after infant cardiac surgery. *Eur J Cardiothorac Surg*. 2009;36:40-7.
6. Gaynor JW, Wernovsky G, Jarvik GP, Bernbaum J, Gerdes M, Zackai E, et al. Patient characteristics are important determinants of neurodevelopmental outcome at one year of age after neonatal and infant cardiac surgery. *J Thorac Cardiovasc Surg*. 2007;133:1344-53.
7. Mahle WT. Neurologic and cognitive outcomes in children with congenital heart disease. *Curr Opin Pediatr*. 2001;13:482-6.
8. Anderson PJ, De Luca CR, Hutchinson E, Roberts G, Doyle LW; Victorian Infant Collaborative. Underestimation of developmental delay by the new Bayley-III scale. *Arch Pediatr Adolesc Med*. 2010;164:352-6.
9. Barach P, Johnson JK, Ahmad A, Galvan C, Bognar A, Duncan R, et al. A prospective observational study of human factors, adverse events and patient

CHD

outcomes in surgery for pediatric cardiac disease. *J Thorac Cardiovasc Surg*. 2008;136:1422-8.
10. Benavidez OJ, Gauvreau K, del Nido P, Bacha E, Jenkins KJ. Complications and risk factors for mortality during congenital heart surgery admissions. *Ann Thorac Surg*. 2007;84:147-55.
11. Larrazabal LA, del Nido PJ, Jenkins KJ, Gauvreau K, Lacro R, Colan S, et al. Measurement of technical performance in congenital heart surgery: a pilot study. *Ann Thorac Surg*. 2007;83:179-84.
12. Bacha EA, Larrazabal LA, Pigula FA, Gauvreau K, Jenkins KJ, Colan SD, et al. Measurement of technical performance in surgery for congenital heart disease: the stage I Norwood procedure. *J Thorac Cardiovasc Surg*. 2008; 136:993-7.
13. Karamichalis JM, Thiagarajan RR, Liu H, Mamic P, Gauvreau K, Bacha EA. Stage I Norwood: optimal technical performance improves outcomes irrespective of pre-operative physiologic status or case-complexity. *J Thorac Cardiovasc Surg*. 2010;139:962-8.
14. Nathan M, Karamichalis J, Liu H, del Nido PJ, Pigula F, Thiagarajan R, et al. Intra-operative adverse events can be compensated in infants after cardiac surgery: a prospective study. *J Thorac Cardiovasc Surg*. 2011;142:1098-107.
15. Karamichalis JM, Colan SD, Nathan M, Pigula FA, Baird C, Marx G, et al. Technical performance scores in congenital cardiac surgery: a quality assessment initiative. *Ann Thorac Surg*. 2012;94:1317-23.
16. Nathan M, Karamichalis JM, Colan S, Emani S, Baird C, Pigula F, et al. Surgical technical performance scores are predictors for late mortality and unplanned reinterventions in infants after cardiac surgery. *J Thorac Cardiovasc Surg*. 2012;144:1095-101.
17. Nathan M, Pigula FA, Liu H, Gauvreau K, Colan SD, Fynn-Thompson F, et al. Inadequate technical performance scores are associated with late mortality and need for late re-intervention in a 13 month cohort of patients followed for 4 years. *Ann Thorac Surg*. 2013;96:664-9.
18. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus based method for risk adjustment for surgery for congenital heart disease. *J Thorac Cardiovasc Surg*. 2002;123:110-8.
19. Jenkin KJ. Risk adjustment for congenital heart surgery: the RACHS-1 method. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2004;7: 180-4.
20. Newburger JW, Sleeper LA, Bellinger DC, Goldberg CS, Tabbutt S, Lu M, et al. Early developmental outcome in children with hypoplastic left heart syndrome and related anomalies: the single ventricle reconstruction trial. *Circulation*. 2012;125:2081-91.
21. Wernovsky G, Newburger J. Neurologic and developmental morbidity in children with complex congenital heart disease. *J Pediatr*. 2003;142:6-8.
22. Wypij D, Newburger JW, Rappaport LA, duPlessis AJ, Jonas RA, Wernovsky G, et al. The effect of duration of deep hypothermic circulatory arrest in infant heart surgery on late neurodevelopment: the Boston Circulatory Arrest Trial. *J Thorac Cardiovasc Surg*. 2003;126:1397-403.
23. Newburger JW, Wypij D, Bellinger DC, du Plessis AJ, Kuban KC, Rappaport LA, et al. Length of stay after infant heart surgery is related to cognitive outcome at age 8 years. *J Pediatr*. 2003;143:67-73.
24. McGrath E, Wypij D, Rappaport LA, Newburger JW, Bellinger DC. Prediction of IQ and achievement at age 8 years from neurodevelopmental status at age 1 year in children with D-transposition of the great arteries. *Pediatrics*. 2004;114: e572-6.
25. Mahle WT, Lu M, Ohye RG, William Gaynor J, Goldberg CS, Sleeper LA, et al. A predictive model for neurodevelopmental outcome after the Norwood procedure. *Pediatr Cardiol*. 2013;34:327-33.

TABLE E1. Univariate analysis of Technical Performance Score classes according to preoperative and postoperative factors

Factor	Class 1 (optimal; n = 83)	Class 2 (adequate; n = 44)	Class 3 (inadequate; n = 13)	*P* value
Preoperative				
Gestational age (wk)	39 (30-42)	39 (35-41)	38 (36-40)	.06
Neonates	43 (52)	16 (36)	9 (69)	.08
RACHS-1 risk category				<.001
1	4 (5)	0 (0)	0 (0)	
2	33 (40)	21 (48)	1 (8)	
3	25 (30)	15 (34)	2 (15)	
4	14 (17)	8 (18)	5 (38)	
6	7 (8)	0 (0)	5 (38)	
Postoperative				
ECMO	5 (6)	2 (5)	6 (46)	<.001
Intubation time (d)	2 (1-94)	2 (1-11)	13 (1-97)	<.001
ICU stay (d)	4 (1-100)	3 (1-36)	16 (2-112)	<.001
Hospital stay (d)	11 (3-111)	9 (3-54)	51 (6-132)	<.001

Data presented as median (range) or n (%). *P* values compared all 3 TPS classes. *RACHS-1,* Risk Adjustment for Congenital Heart Surgery; *ECMO,* extracorporeal membrane oxygenation; *ICU,* intensive care unit; *TPS,* Technical Performance Score.

CHD

TABLE E2. Univariate analysis of Bayley Scale of Infant Development cognitive composite score according to preoperative and postoperative factors

Factor	Patients (n)	Mean ± SD	*P* value
Chromosomal anomaly			.001
Present	11	89 ± 10	
Absent	129	101 ± 18	
Nonchromosomal anomaly			.08
Present	18	93 ± 19	
Absent	122	101 ± 17	
RACHS-1 category			.17
1-2	59	103 ± 16	
3	42	99 ± 19	
4	27	99 ± 13	
6	12	92 ± 24	
RACHS-1 category grouping			.07
1-4	128	101 ± 17	
6	12	92 ± 24	
Need for ECMO			<.001
Yes	13	87 ± 20	
No	127	102 ± 17	
Feeding status at discharge			.007
Oral	132	102 ± 17	
Not oral	8	79 ± 17	
Statistics		r_s	
Intubation time (d)		−0.17	.05
ICU stay (d)		−0.14	.10
Postoperative LOS (d)		−0.13	.14
Hospital LOS (d)		−0.14	.10

No significant associations were found with gender, gestational age, age at surgery, weight at surgery, use of a pump, or palliated circulation (all $P > .20$). *SD,* Standard deviation; *RACHS-1,* Risk Adjustment for Congenital Heart Surgery; *ECMO,* extracorporeal membrane oxygenation; *ICU,* intensive care unit; *LOS,* length of stay.

TABLE E3. Univariate analysis of Bayley Scale of Infant Development motor composite score according to preoperative and postoperative factors

Factor	Patients (n)	Mean ± SD	*P* value
Chromosomal anomalies			.007
Yes	11	78 ± 12	
No	127	90 ± 17	
Nonchromosomal anomalies			.19
Yes	18	84 ± 17	
No	120	90 ± 17	
RACHS-1 category			.08
1-2	58	93 ± 16	
3	42	88 ± 17	
4	26	87 ± 14	
6	12	82 ± 17	
RACHS-1 category grouping			.10
1-4	126	90 ± 16	
6	12	82 ± 17	
Ventricles			.03
SV	28	83 ± 17	
BiV	110	91 ± 16	
Circulatory arrest used			.06
Yes	31	84 ± 16	
No	107	91 ± 17	
Need for ECMO			<.001
Yes	13	69 ± 17	
No	125	91 ± 15	
Oral feeding status at discharge			.008
Yes	130	91 ± 16	
No	8	68 ± 18	
Statistics		r_s	
Intubation time (d)		−0.31	<.001
ICU stay (d)		−0.26	.002
Postoperative LOS (d)		−0.27	.002
Hospital LOS (d)		−0.27	.001

No significant associations were found with gender, gestational age, age at surgery, weight at surgery, or use of a pump (all $P > .20$). *SD,* Standard deviation; *RACHS-1,* Risk Adjustment for Congenital Heart Surgery; *SV,* single ventricle; *BiV,* biventricular repair; *ECMO,* extracorporeal membrane oxygenation; *ICU,* intensive care unit; *LOS,* length of stay.

CHD

TABLE E4. Multivariate analysis of Bayley Scale of Infant Development cognitive composite score adjusting for preoperative factors (excluding chromosomal anomalies Analysis; total $R^2 = 11.1\%$)

	Preoperative risk factors*		Preoperative risk factors plus TPS		
Factor	Regression coefficient ± SE	*P* value	Regression coefficient ± SE	*P* value	Partial R^2 (%)
Nonchromosomal anomalies	−10.4 ± 4.3	.02	−10.4 ± 4.3	.02	4.4
RACHS-1 category 6	−11.4 ± 5.2	.03	−7.5 ± 5.5	.18	1.5
TPS class (vs optimal)					4.2
Adequate (class 2)	—		−1.7 ± 3.4	.62	
Inadequate (class 3)	—		−12.5 ± 5.5	.03	

TPS, Technical Performance Score; *SE,* standard error; *RACHS-1,* Risk Adjustment for Congenital Heart Surgery. *Significant at the $P = .10$ level; in the model on the right (with TPS), we see that an inadequate TPS remained significantly associated with a lower score after adjusting for chromosomal and nonchromosomal anomalies and RACHS-1 risk category.

TABLE E5. Multivariate analysis of Bayley Scale of Infant Development motor composite score adjusting for preoperative factors (excluding chromosomal anomalies; total $R^2 = 12\%$)

Factor	Preoperative and surgical risk factors*		Preoperative and surgical risk factors plus TPS		
	Regression coefficient ± SE	*P* value	Regression coefficient ± SE	*P* value	Partial R^2 (%)
Nonchromosomal anomalies	−8.0 ± 4.1	.05	−7.9 ± 4.1	.06	2.9
Single ventricle	−7.4 ± 3.6	.05	−6.4 ± 3.8	.11	2.4
Circulatory arrest	−7.1 ± 3.4	.04	−6.2 ± 3.5	.08	2.5
TPS class (vs optimal)					1.9
Adequate (class 2)	—		−1.4 ± 3.3	.68	
Inadequate (class 3)	—		−7.6 ± 5.2	.15	

TPS, Technical Performance Score; *SE,* standard error. *Significant at the $P = .10$ level; in the model on the right (with TPS), we see that an inadequate TPS had only a borderline association with a lower score after adjusting for chromosomal and nonchromosomal anomalies, single ventricle palliation, and the use of circulatory arrest.

TABLE E6. Sensitivity analysis comparing patients who did and did not undergo neurodevelopmental testing

Variable	ND testing (n = 140)	No ND testing (n = 257)	*P* value
Gender			.52
Male	77 (55)	144 (59)	
Female	63 (45)	101 (41)	
Gestational age (wk)	39 (30-42)	39 (28-41)	.36
Age at surgery (d)	34 (1-333)	24 (1-359)	.81
Weight at surgery (kg)			.31
<5	100 (71)	162 (66)	
≥5	40 (29)	83 (34)	
Chromosomal anomaly			.27
Yes	11 (8)	12 (5)	
No	129 (29)	233 (95)	
Nonchromosomal anomaly			.009
Yes	18 (13)	12 (5)	
No	122 (87)	233 (95)	
RACHS-1 risk category			.09
1	4 (3)	5 (2)	
2	55 (39)	87 (36)	
3	42 (30)	58 (24)	
4	27 (19)	62 (25)	
6	12 (9)	22 (9)	
7	0 (0)	11 (4)	
Pump use			.16
Yes	122 (87)	225 (92)	
No	18 (13)	20 (8)	
Circulatory arrest time (min) (n = 32 and n = 82)	16 (2-87)	29 (1-99)	.08
Ventricles			1.0
Single ventricle/palliated	28 (20)	48 (20)	
Biventricular repair	112 (80)	197 (80)	
TPS			.77
Class 1 (optimal)	83 (59)	140 (57)	
Class 2 (adequate)	44 (31)	73 (30)	
Class 3 (inadequate)	13 (9)	31 (13)	
Class 4 (TPS score not assigned)	0 (0)	1 (<1)	

Data presented as n (%) or median (range). The patients who had undergone ND testing did not differ significantly from the group of patients who had not undergone ND testing within the same period, except for a greater rate of nonchromosomal anomalies in those who had undergone testing. RACHS-1 risk category 7 represents those patients for whom a RACHS-1 category could not be assigned. *ND,* Neurodevelopmental; *RACHS-1,* Risk Adjustment for Congenital Heart Surgery; *TPS,* Technical Performance Score.

CHD