

Neurodevelopmental Outcomes in Preschool Survivors of the Fontan Procedure

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Neurodevelopmental dysfunction is the most common, and potentially most disabling, long-term complication of congenital heart defects and their treatment.

There has been a general perception that early ND outcomes for patients with functional single ventricle are significantly worse than for children with other types of CHD.

Study Design

- Prospective study evaluating the association between ND dysfunction and apolipoprotein E (*APOE*) genotype in neonates and infants undergoing surgery for CHD
- Detailed neurobehavioral evaluation at 4 years age
- Compare ND outcomes of preschool survivors of the Fontan procedure with those of children with CHD undergoing biventricular repair
- Investigate predictors of ND outcome for patients with single ventricle CHD, including HLHS.

Study Population

- Between September 1998 and April 2003, 675 eligible infants underwent cardiac surgery and 550 (81%) were enrolled
- 486 patients were alive and eligible for the 4-year neurodevelopmental evaluation
 - *381 patients completed the 4 year evaluation (78% of eligible)*
- Only significant difference in baseline characteristics between returning and non-returning patients was under-representation of African-Americans in the returning patients (21% vs. 29%)
- Genetic evaluation normal in 269/381 (71%)

Study Population

- Patients were considered for enrollment in the current analysis if they had achieved an end-state of either a completed BV repair or a Fontan operation.
- Patients who had not achieved one of these end-states or who had undergone cardiac transplantation were not considered in this analysis.
- A total of 365 patients met entry criteria for this analysis. BV repair had been achieved in 253 patients and 112 patients had undergone the Fontan procedure.

Baseline Characteristics

No differences between SV and BV patients for:

gender

ethnicity

gestational age

APOE genotype

maternal education

socioeconomic status

Definite genetic anomalies more common in BV patients ($p < 0.01$)

Preoperative mechanical ventilation more common in SV patients
($p < 0.01$)

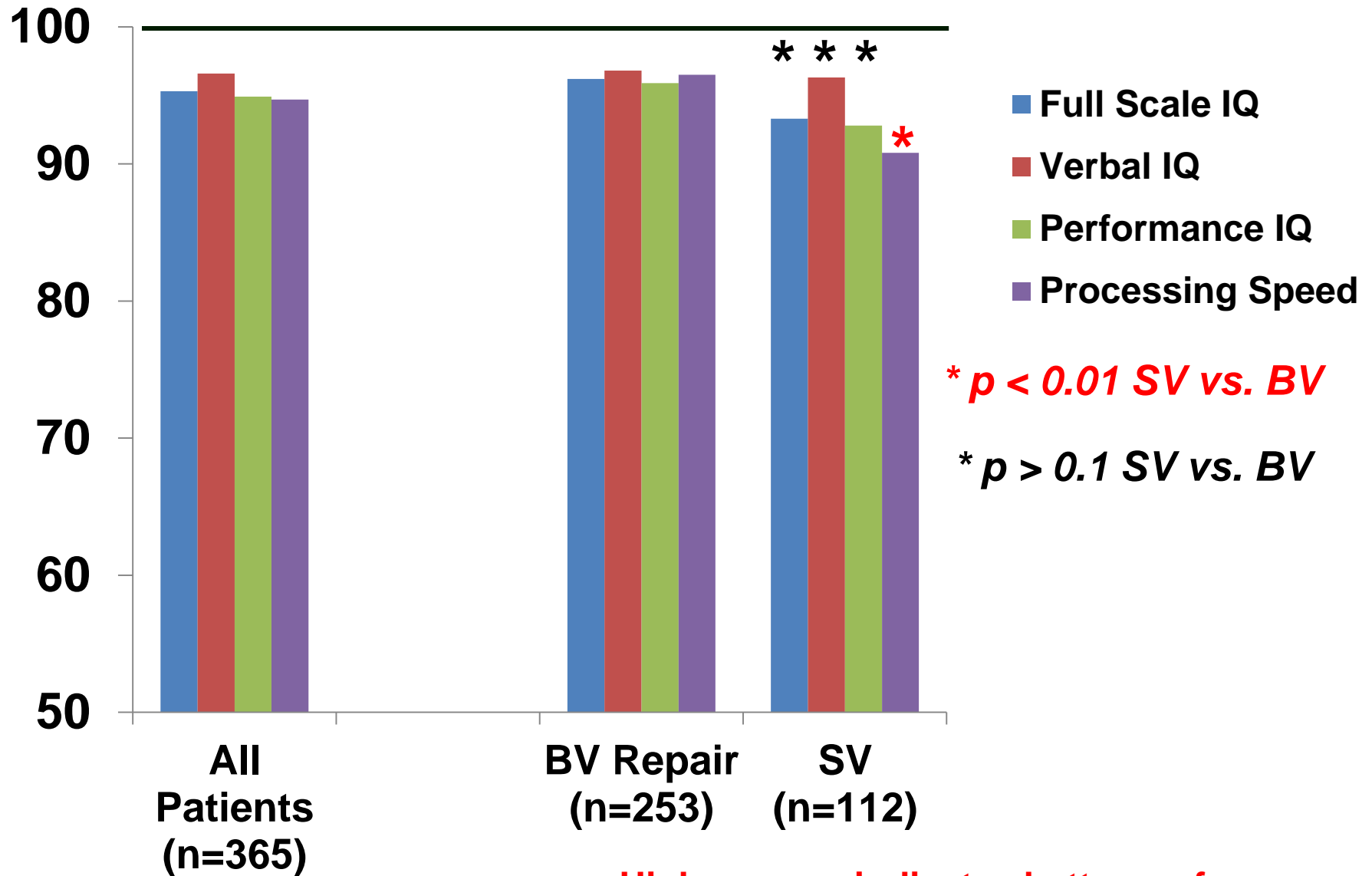
Operative Management

Variable	BV (n = 253)	SV (n = 112)	p-value
<i>First Operation</i>			
Age (days)	55.4 ± 56.7	17.1 ± 37.2	< 0.01
Weight (kilograms)	4.1 ± 1.3	3.5 ± 1.0	< 0.01
CPB Time (min)	70.9 ± 42.9	53.6 ± 25.8	< 0.01
DHCA Used (Yes)	112 (44%)	99 (88%)	< 0.01
DHCA Time (min)	35.8 ± 16.4	42.5 ± 15.8	< 0.01
Hematocrit after Hemodilution (%)	27.4 ± 4.1	28.8 ± 3.9	0.01
Postoperative Length of Stay	10.7 ± 13.2	14.4 ± 11.3	< 0.01

Operative Management

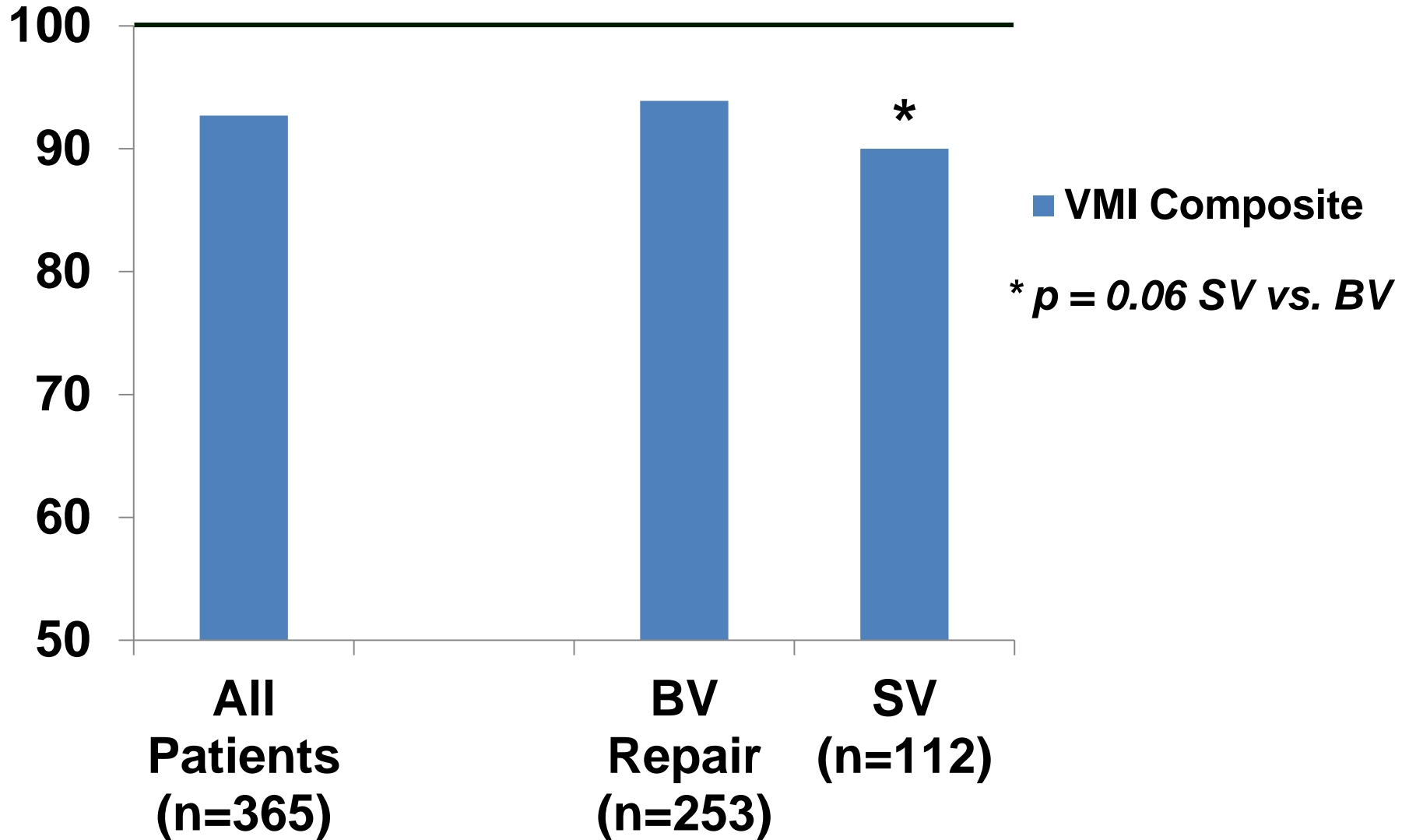
Variable	BV (n = 253)	SV (n = 112)	p-value
Additional Operations with CPB (Yes)	28 (11%)	112 (100%)	< 0.01
Mean Additional Operations with CPB	0.1 ± 0.4	1.9 ± 0.3	< 0.01
Additional CPB Time (min)	68.6 ± 50.7	108.4 ± 62.8	0.02
Additional DHCA (Yes)	7 (3%)	98 (88%)	< 0.01
Additional DHCA Time (min)	22.3 ± 13.3	46.7 ± 22.3	< 0.01

Cognition



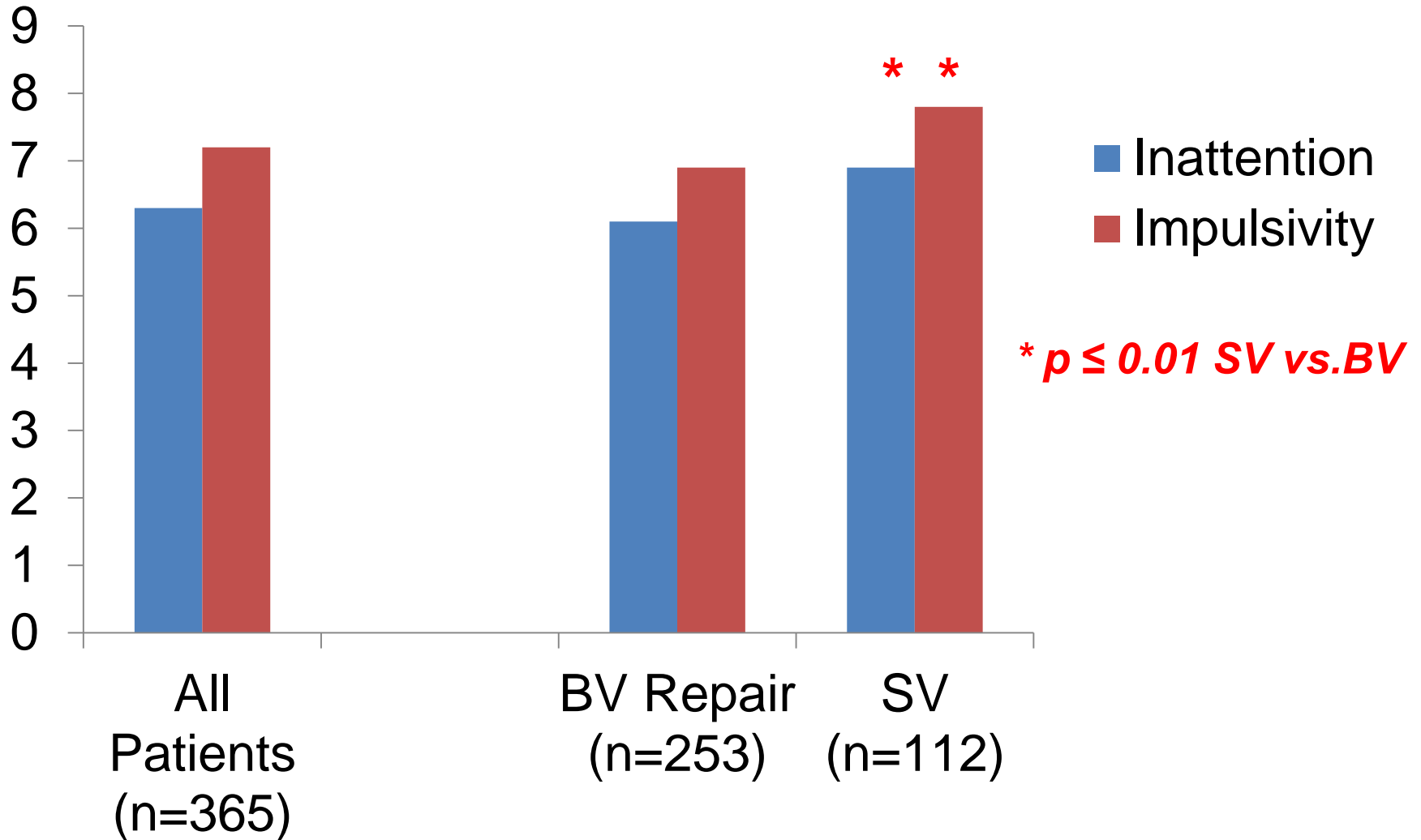
Higher score indicates better performance

Visual-Motor Integration



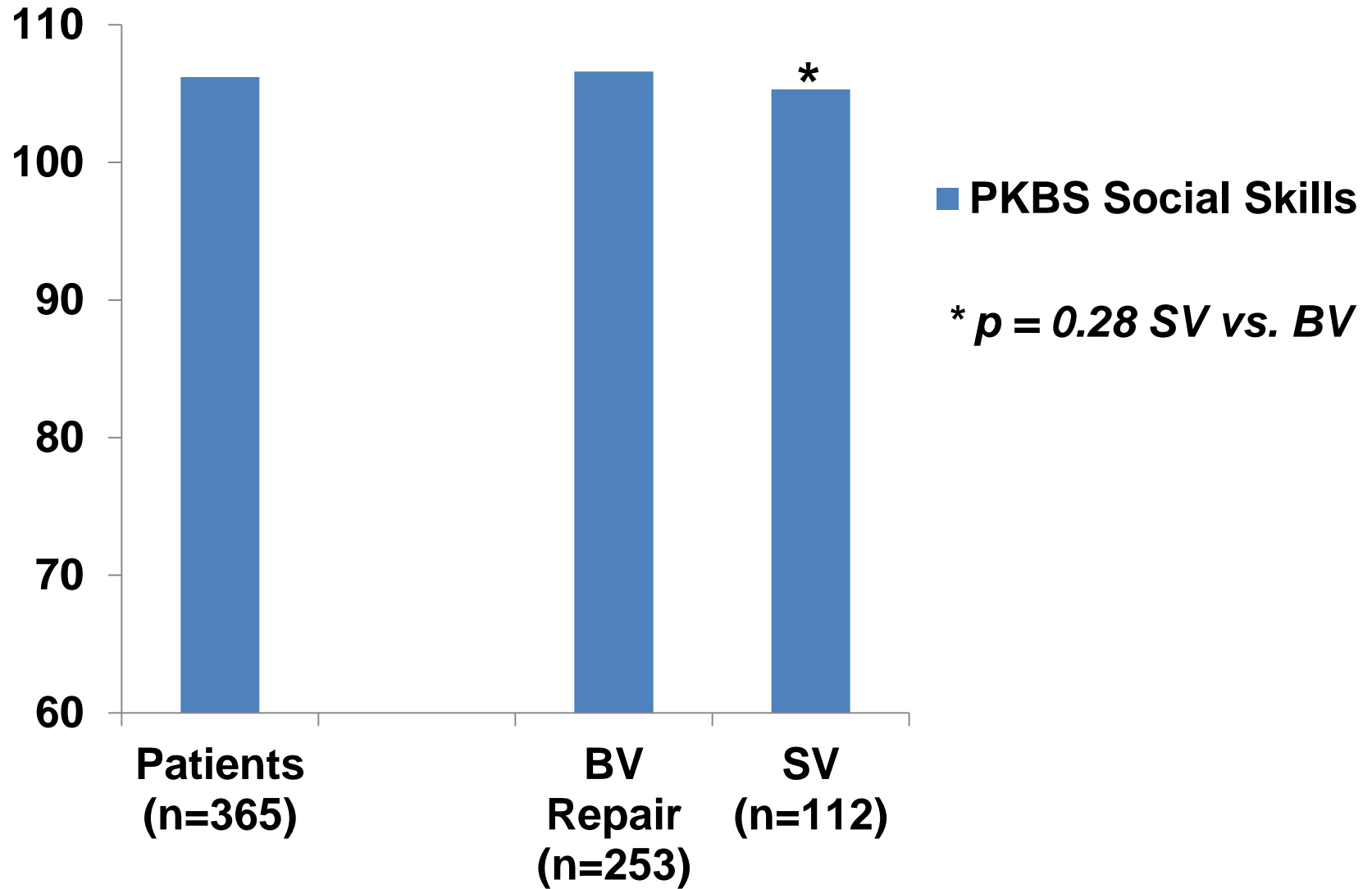
Higher score indicates better performance

Inattention and Impulsivity



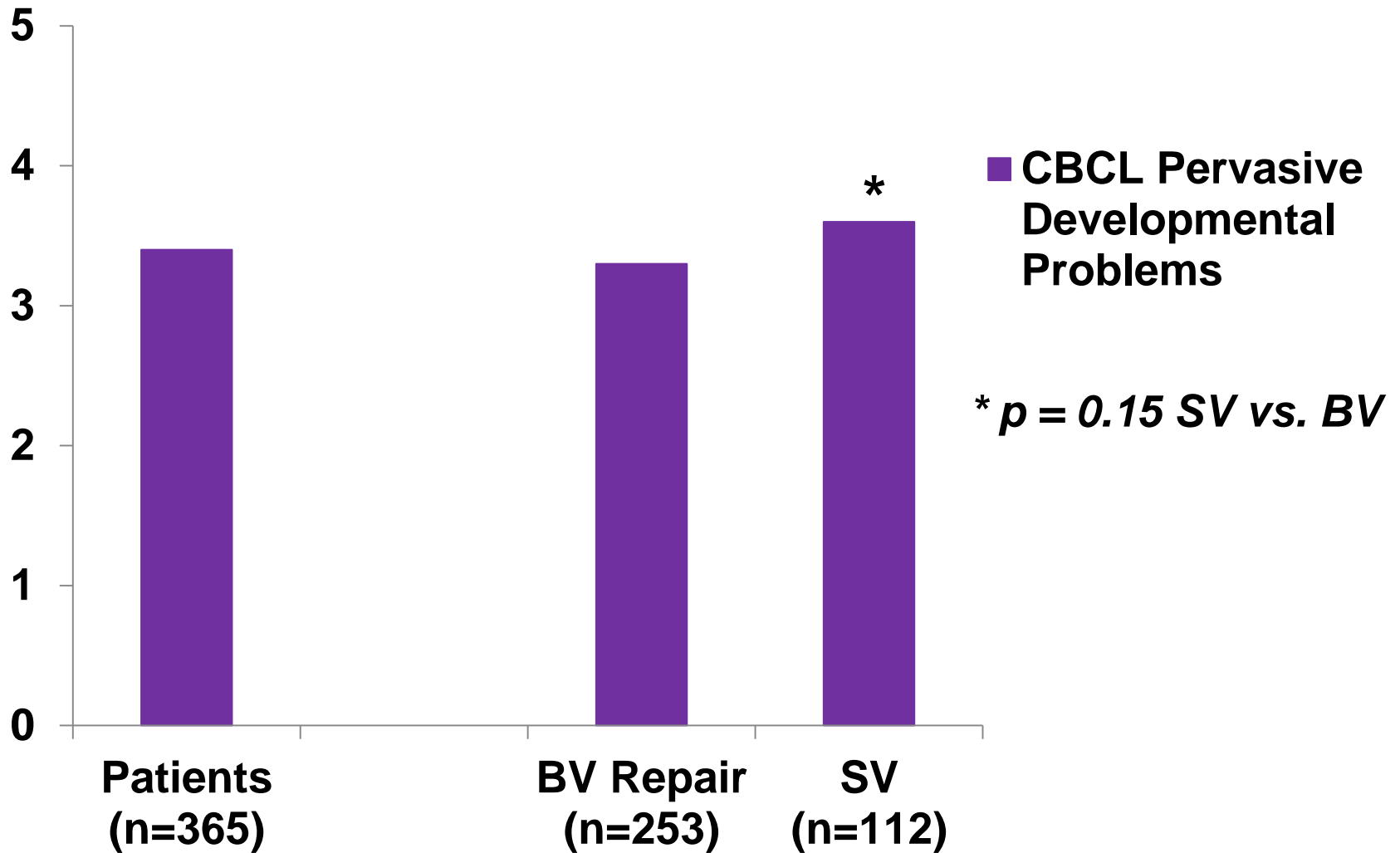
Higher score indicates worse performance

Social Skills



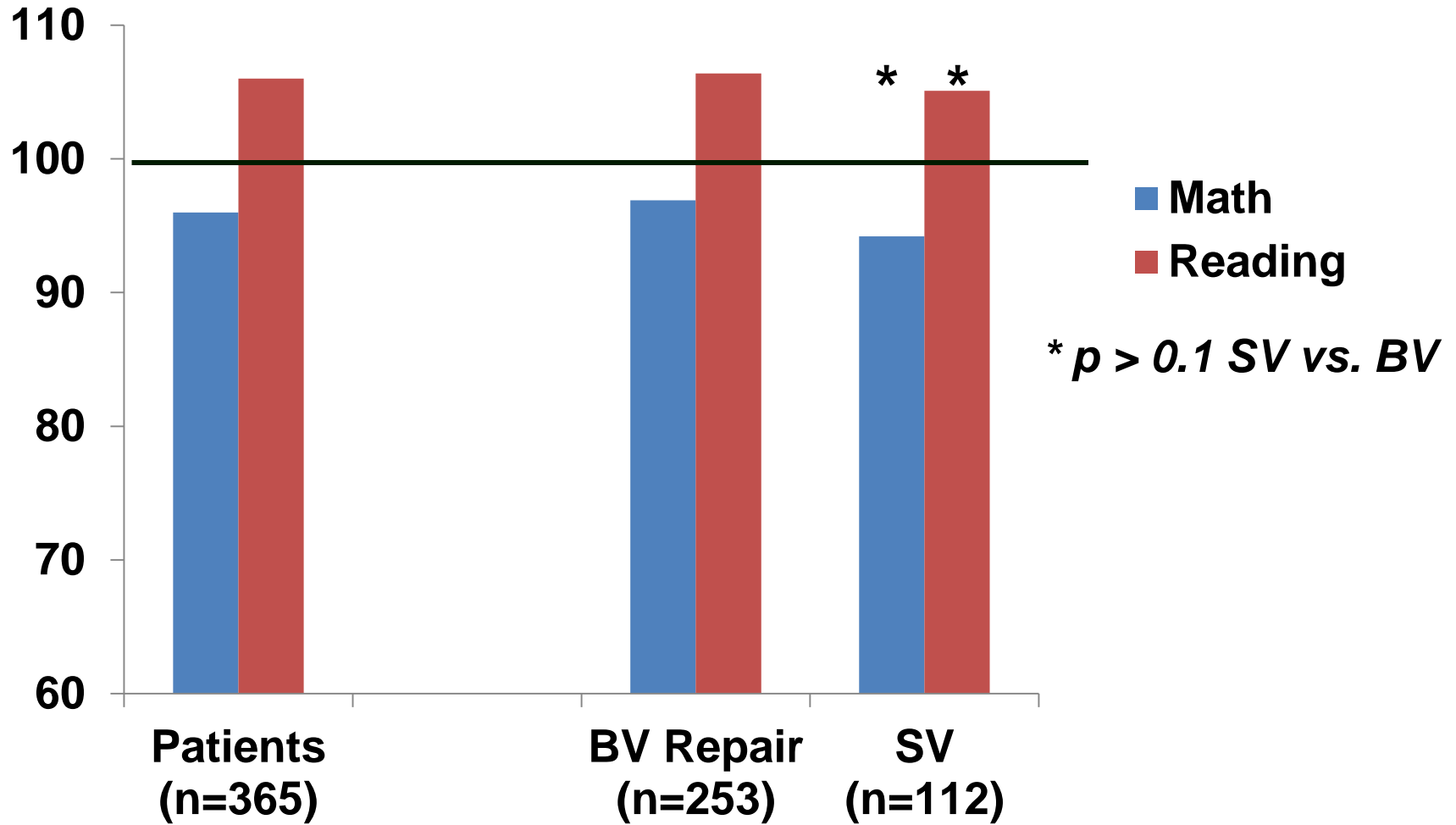
Higher score indicates better performance

Restricted Behaviors



Higher score indicates worse performance

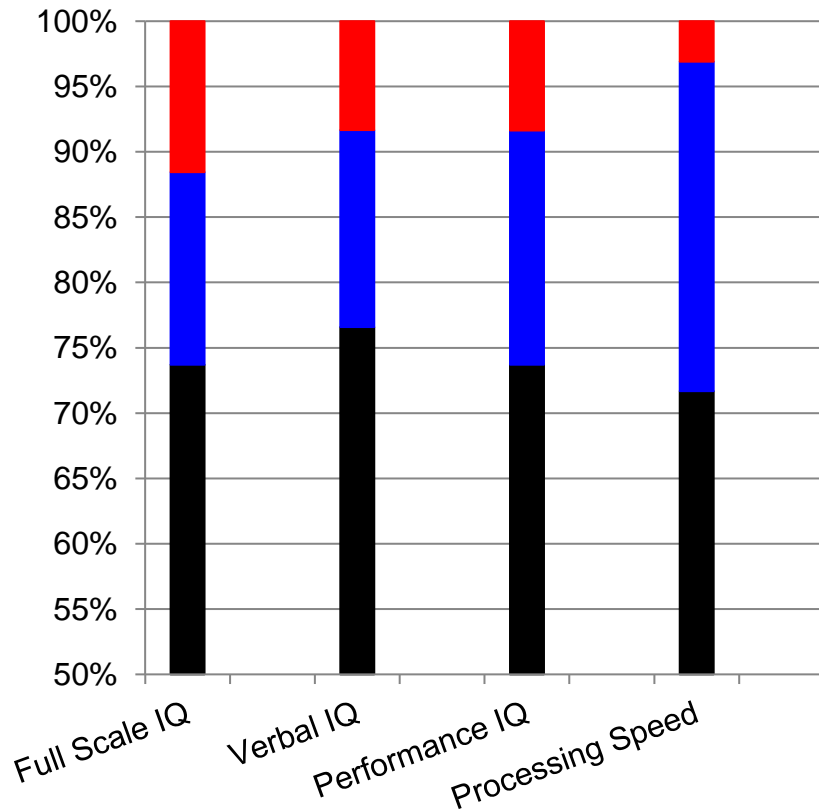
Academic Achievement



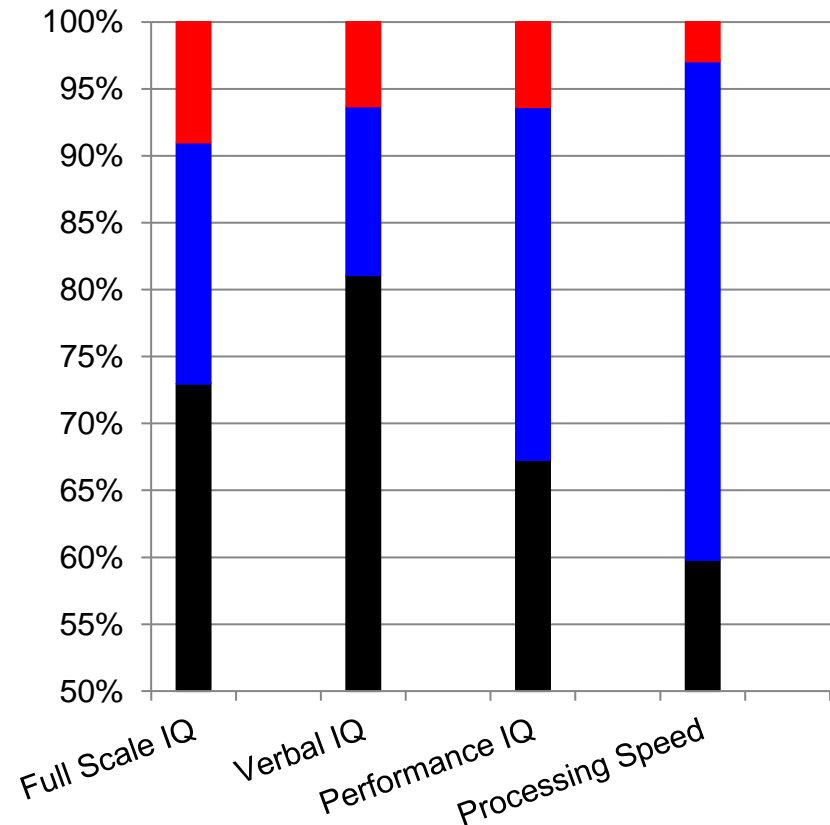
Higher score indicates better performance

Severity of Impairment

Biventricular Repair



Single Ventricle



Normal: < 1 SD below mean

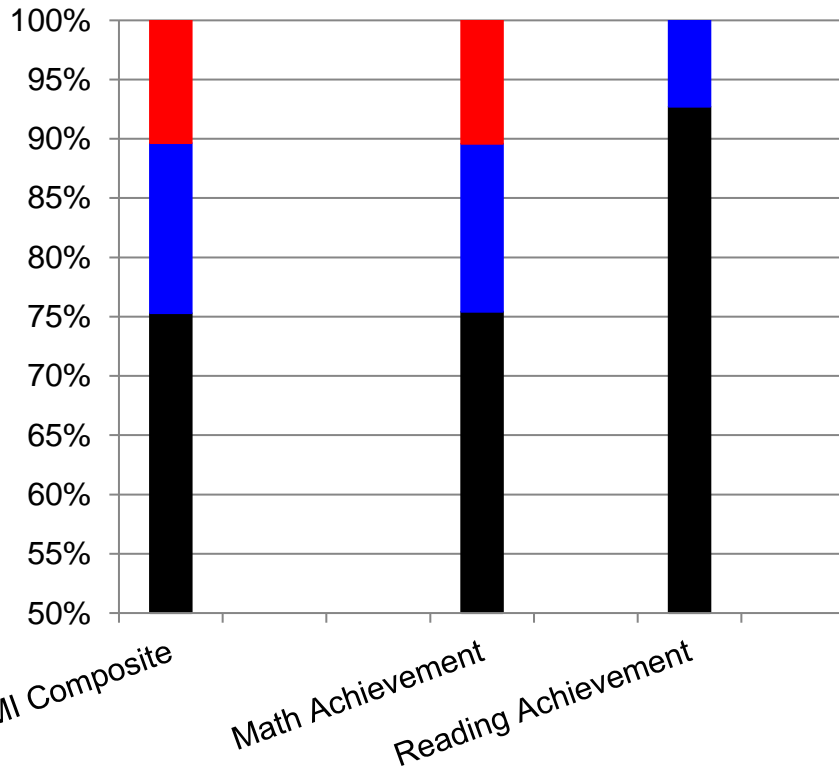
Moderate Impairment: 1-2 SD below mean

Severe impairment: > 2 SD below mean

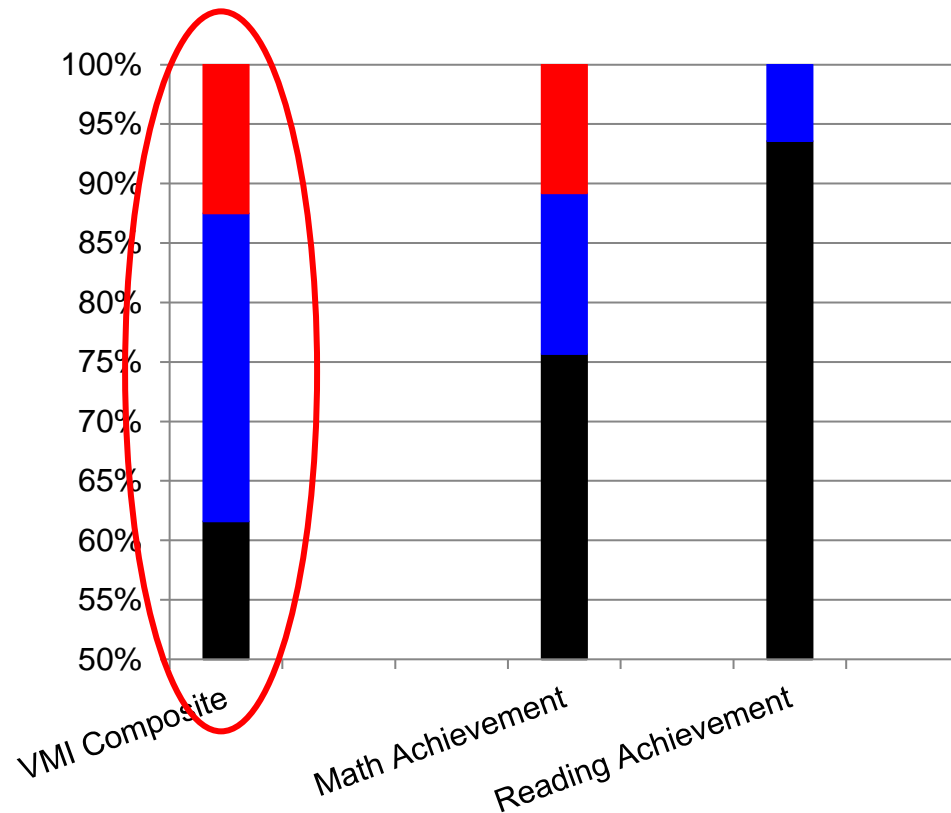
p > 0.05 for all

Severity of Impairment

Biventricular Repair



Single Ventricle



Normal: < 1 SD below mean

Moderate Impairment: 1-2 SD below mean

Severe impairment: > 2 SD below mean

p > 0.5 for math and reading

p = 0.02 for VMI

Predictors of ND Outcomes after the Fontan Procedure

Cognition:

Larger birth weight and older gestational age associated with better performance

Higher HCT associated with better performance

Longer post-operative LOS associated with worse performance

HLHS and use of DHCA not associated with worse performance

Predictors of ND Outcomes after the Fontan Procedure

Behavior and Social Skills:

Larger birth head circumference and older gestational age associated with better performance

Preoperative mechanical ventilation associated with worse performance

Genetic anomalies and lower SES associated with worse performance

HLHS and use of DHCA not associated with worse performance

Predictors of ND Outcomes after the Fontan Procedure

Academic Achievement:

Greater maternal education associated with better performance

Longer post-operative LOS associated with worse performance

**HLHS and use of DHCA not
associated with worse performance**

Conclusions

- Unadjusted ND outcomes for preschool aged survivors of the Fontan procedure are similar to those for children with CHD undergoing BV repair for most domains.
- Among the Fontan patients, HLHS was not associated with worse outcomes compared to other forms of SV.
- Most children function within the low normal range for the domains tested. The occurrence of severe impairment is greater compared to the general population, but is present in < 10% of patients for most domains tested.