Use of Early Remedial Services in Children with Transposition of the Great Arteries

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Objectives To characterize the prevalence of use of early remedial services and its associated demographic, medical, and cognitive factors in children aged 4-6 years with corrected transposition of the great arteries (TGA).
Study design This was a prospective study of neurocognitive outcomes after TGA. Children underwent formal neuropsychological testing including general intelligence and a comprehensive battery of executive functions (EF) including motor and interference control, short-term memory, and working memory as well as cognitive flexibility. Parental reports on the children's behavior and EF were also evaluated. Demographic factors and preoperative, intraoperative, and postoperative factors as well as cognitive factors were examined according to the current use of remediation.
Results Forty-five patients (67% male) and their parents participated in this study. Twenty-four (53%) patients were receiving remedial services. Male sex, a postnatal diagnosis of TGA, and a longer postoperative intensive care unit stay were significantly associated with use of remediation. Children receiving remediation had lower EF scores, had more severe EF deficits as observed by formal testing, and were rated as having more behavioral daily life difficulties. However, in the group without remediation, 13 children (43%) also displayed EF deficits rated as moderate to severe.
Conclusions Demographic and medical factors could help identify children at higher risk for neurocognitive delays. Evaluation of executive functioning from an early age may influence referral for remediation. *(J Pediatr 2013;163:1105-10)*.

Progress in survival rates and physical outcomes has led to the important recognition of neurobehavioral and cognitive impairments for children with congenital heart disease (CHD). The most recent studies suggest the presence of a distinctive neurodevelopmental pattern of dysfunction including mild motor deficits, impaired social interaction and communication skills, inattention, and deficits in executive functions (EF).[1-4] These impairments can have detrimental repercussions on academic achievement, psychosocial development, and ultimately quality of life.[1] It has been reported that nearly one-half of school-aged children with CHD were receiving some form of educational service[5] and that 65% of adolescents with transposition of the great arteries (TGA) had a frequent history of special services use.[6] However, very few studies have addressed this question systematically, and only 1 study focused on the pattern and characteristics of remedial service use, with only 23% benefiting from services.[7]

The prevalence and predictors of the use of remediation use at an earlier age where cognitive and behavioral impairments are emerging such as during the preschool years remain unknown. Children with CHD including TGA display significant behavioral and EF dysfunction[2-4,8] observable even before entry to primary school.[9] A high prevalence of hyperactivity/impulsivity symptoms consistent with attention-deficit hyperactivity disorder (ADHD)[5] has also been reported. EF including motor and interference control, short-term memory, and working memory as well as cognitive flexibility undergo crucial improvements in the preschool period and are important precursors of children's behavioral and social adaptation.[10-12] These higher-order cognitive skills are strong predictors of later academic achievement including verbal and math learning in children.[10-17] It has been shown that early interventions in children with cognitive disability promote greater achievement.[18] The aim of this study was to characterize the prevalence of use of remedial services and associated factors in children with TGA. We sought to determine the association of demographic and pre-, intra-, and postoperative factors as well as cognitive outcomes to the use of early remedial services.

Methods

All neonates born with TGA between 2003 and 2005 and operated at the referral center for complex congenital heart malformations, Necker Children's Hospital

ADHD	Attention-deficit hyperactivity disorder
CHD	Congenital heart disease
EF	Executive functions
ICU	Intensive care unit
NEPSY	Developmental Neuropsychological Assessment
TGA	Transposition of the great arteries

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(Paris, France), were screened for participation in a single center prospective study of neurocognitive outcomes. Patients meeting inclusion criteria received a diagnosis of TGA with intact ventricular septum or ventricular septal defect and underwent a neonatal arterial switch operation under continuous full-flow cardiopulmonary bypass. Neonates with associated genetic syndromes (including 22q11 deletion), associated extra-cardiac pathologies or cardiovascular anomalies requiring aortic arch reconstruction, and the use of deep hypothermic circulatory arrest or additional open surgical procedures as well as neonates with a birth weight less than 2.5 kg were excluded. Children whose native language was not French and/or who were living outside the Paris area at the time of enrollment were also excluded from participation. Data from a control group of same aged children (n = 45) living in the same geographic area was used as a reference to compare the prevalence of remedial service use.

The institutional ethics committee of Necker Children's Hospital approved the protocol and informed consent. Eligible families were contacted by a letter explaining the aim of the study with telephone follow-up to further explain the protocol. Families who agreed to participate were scheduled for a 2 to 2.5-hour visit at the hospital, and written informed consent was obtained from the parent(s) and the child. A cognitive evaluation was performed on all children by a trained neurodevelopmental psychologist who did not review medical records before conducting the neuropsychological battery.

Cognitive assessment included a nonverbal IQ evaluation using the Columbia Mental Maturity Scale.[19] Motor control was assessed with the knock and tap subtest from the Developmental Neuropsychological Assessment (NEPSY)[20]; interference control and impulse control were measured with the Animal Stroop test,[21] a pictorial version of the Stroop task that does not require reading abilities. This test provides an evaluation of interference sensitivity and control of attention expressed by reaction times as well as an evaluation of cognitive impulse control skills expressed by the total number of incorrect responses. Verbal short-term memory and verbal working memory were assessed with the forward and backwards digit span subtests from the *Wechsler Intelligence Scale for Children-fourth edition*.[22] Spatial immediate memory and spatial working memory were assessed with the spatial span task.[23] In this test, analogous to the verbal working memory task, the stimuli are visual (a set of small squares randomly positioned). Children reproduce a sequence of squares locations in the same order as they saw it and then backwards. Finally, cognitive flexibility was evaluated with the Dimensional Card Sorting Test,[24] a well-known neuropsychological test for preschool children. Normative values for all tests were obtained from national standardizations and previous references published.[2,9,21] Parental reports on the children's behavior and EF were obtained using the Behavior Rating Inventory of Executive Function-preschool and school-age versions[25] for children between 4 and 6 years 11 months. For this scale we analyzed the global executive composite score, a T score (expected mean = 50; SD = 10) with a higher T score indicating greater impairment (**Appendix**; available at www.jpeds.com).

Information regarding remedial services use was gathered through parental reports during the visits. A short written survey with a multiple-choice format was used to collect diverse data regarding health status, demographic variables, and developmental information including current remedial service use. Patient's medical records were reviewed for potential risk factors associated with cognitive deficits and remedial service use. Preoperative medical factors included pre- vs postnatal diagnosis of TGA, presence or absence of a ventricular septal defect, birth weight, gestational age, Apgar score at 5 minutes, hemodynamic condition (metabolic acidosis defined with a pH level <7.2), and the need for balloon-atrial septostomy. Age at the arterial switch operation, total bypass time and total cross-clamp time, and postoperative intensive care unit (ICU) stay duration were also examined. Demographic variables including sex, age at evaluation, family socioeconomic status, and parental educational levels were also recorded.

Statistical Analyses

Statistical analyses were performed with Statistica software (v. 9.1; StatSoft Inc, Maisons-Alfort, France). Descriptive results are presented with means (SD) and proportions in each group. Demographic and medical variables were compared between groups by using χ^2 tests for categorical variables and ANOVA for numerical variables. Differences in cognitive scores were assessed using ANOVA with post-hoc testing (Tukey Honestly Significant Difference Test) for specific comparisons across groups for all continuous variables. Multivariate logistic regression analyses including separate analyses with demographic and medical variables were performed to identify significant predictors of remedial service use. Scores in each EF subtest were compared with normative values and deficits were described according to their severity as "mild" (−1 SD) or "moderate to severe" (−2 SD or below). The total number of mild and moderate to severe deficits were calculated for each individual, each one ranging from 0 = no deficits to 8 = deficits in all EF subtests. Mean numbers of mild and moderate to severe deficits were then calculated for the groups (with and without remediation), and Poisson regression analyses were used for comparisons between the groups. Multiple logistic regression analyses were used to determine significant determinants (medical and demographic) once the severity of EF deficits was adjusted. A P value of .05 was considered significant.

Results

A total of 60 eligible children born between 2003 and 2005 were identified in the database. Six families could not be contacted because of a change of address. Fifty-four families were contacted for enrollment in a single-center prospective study of neurocognitive outcomes. Parents of 6 children declined