Intellectual, neuropsychological, and behavioral functioning in children with tetralogy of Fallot

Marijke Miatton, Dpsych,[a] Daniël De Wolf, MD, PhD,[b] Katrien François, MD,[c] Evert Thiery, MD,[d] and Guy Vingerhoets, PhD[a]

Objective: Although it is known that pediatric cardiac surgery holds risks for later development, few studies investigated the long-term development in children with tetralogy of Fallot. The purpose of this study was to define their intellectual capacities, neuropsychological profile, and behavioral functioning 6 to 12 years postoperatively.

Methods: Patients (n = 18; age, 8 years, 3 months ± 1 year, 6 months) were examined with a short-form intelligence scale (Wechsler Intelligence Scale for Children, 3rd edition, Dutch version) and a neuropsychological assessment battery (NEPSY). Their parents completed a behavioral questionnaire. The patient group was compared with an acyanotic congenital heart disease group and a healthy control group.

Results: No significant differences between the patient group and the acyanotic group emerged. Compared with the healthy control group, children with tetralogy of Fallot showed significantly lower scores on the estimated Full Scale IQ ($P < .05$) and on the NEPSY domains Language ($P < .01$) and Sensorimotor Functioning ($P < .01$). Also, the subtests Tower ($P < .05$), Memory for Names ($P < .05$), Narrative Memory ($P < .05$), and Design Copy ($P < .05$) elicited group differences. Parental reports revealed significantly higher scores on attention problems ($P < .05$) and the total problem scale ($P < .05$), as well as significantly lower school performances than those of healthy peers ($P < .01$).

Conclusions: In children with tetralogy of Fallot, we identified a lower estimated full-scale intelligence than in healthy peers and a neuropsychological profile characterized by primarily mild motor deficits and difficulties with language tasks. Parents of the children with tetralogy of Fallot indicated attention problems and rated the child's school competencies to be lower than in healthy control subjects.

From the Laboratory for Neuropsychology, Department of Internal Medicine[a]; Paediatric Cardiology, Department of Paediatrics[b]; Paediatric Cardiac Surgery, Department of Surgery[c]; and Reference Centre for Refractory Epilepsy,[d] Ghent University, Ghent, Belgium.

Received for publication Aug 22, 2006; revisions received Sept 29, 2006; accepted for publication Oct 10, 2006.

Address for reprints: Miatton Marijke, Dpsych, Laboratory for Neuropsychology, Ghent University, De Pintelaan 185, 4 K 3, B-9000 Ghent, Belgium (E-mail: marijke.miatton@ugent.be).

J Thorac Cardiovasc Surg 2007;133:449-55

0022-5223/$32.00

doi:10.1016/j.jtcvs.2006.10.006

Tetralogy of Fallot (TOF) is one of the most common forms of cyanotic congenital heart disease (CHD).[1] A cyanotic defect results in abnormal blood flow through the lungs preventing full oxygenation of the blood and causes symptoms, such as cyanosis, breathlessness, and fatigue, and growth retardation.[2] Today, these children are operated on at an early age to normalize their cardiopulmonary status as soon as possible. Many studies have sought to define differences in functional outcome between cyanotic and acyanotic forms of CHD. In an acyanotic type of CHD, the circulation to the lungs is normal, and there is full oxygenation of the systemic blood. Generally, it is postulated that because of the difference in severity of symptoms, cyanotic forms of CHD result in lower functional outcome than acyanotic forms of CHD.[3] However, this is not always supported by clinical evidence.[4,5]

Only a few studies have engaged in defining the functional outcome in isolated diagnostic groups. Research on children with transposition of the great arteries

Abbreviations and Acronyms

CBCL = Child Behavior Checklist
CHD = congenital heart disease
NEPSY = developmental neuropsychological assessment
SD = standard deviation
TGA = transposition of the great arteries
TOF = tetralogy of Fallot
VSD = ventricular septal defect

(TGA) or TOF, 2 cyanotic forms of CHD, revealed significantly lower scores on academic skills, such as reading, spelling, and arithmetic, compared with those in an acyanotic group.[1] Differences between both cyanotic groups (TGA and TOF) could not be found.[4] Studies on children with TOF showed normal intellectual functioning[5,6] but marked motor dysfunctions and a higher incidence of language deficits.[6] Neuropsychological assessment on adult patients with TOF revealed impairment in executive functioning. These patients also reported lower academic levels, despite having spent more time in school.[7]

Studies on behavior in isolated groups of children with TOF are rare. Moreover, results on behavioral functioning in children with various CHDs are inconsistent. Although some studies report the presence of significantly higher behavioral problem scores in children with CHD,[8,9] other studies conclude that no behavioral problems are present, and sometimes the parents even indicate fewer symptoms than parents of healthy children.[1,10]

Obviously, the division between cyanotic and acyanotic forms of CHD elicits conflicting results. As a consequence, research on separate diagnostic groups might result in the specification of functional outcome according to diagnosis. In addition, although cognitive dysfunctions at adult age and school problems have been mentioned, the neuropsychological profile of children with TOF remains unknown.

The purpose of this study was to define the intellectual capacities, neuropsychological profile, and behavioral functioning of full-time school-attending children with TOF 6 to 12 years postoperatively to identify shortcomings or relative difficulties that can lead to tailored interventional programs. We compared the TOF children with an acyanotic group and with a healthy control group.

Patients and Methods

Patient Characteristics and Medical Data

Patients with various CHDs operated on at Ghent University Hospital between 1995 and 1999 with a birth weight of greater than 2000 g, without perinatal problems, and without noncardiac malformations or genetic abnormalities (Down syndrome, velocardiofacial syndrome, and Di George syndrome) were contacted and invited to participate in the study. From this total group, we selected the children with TOF (n = 29) and matched them with a group of children with an acyanotic CHD. All children with TOF and acyanotic children showing characteristic features of genetic abnormalities at birth had a genetic screening (fluorescence *in situ* hybridization). After exclusion of children with genetic abnormalities (8 children in the TOF group and no children in the acyanotic CHD group), we contacted 21 patients with TOF. One child could not be included because of a severe hearing disorder, and 2 other parents thought participation would be too time consuming. We included 18 patients with TOF (10 boys; age, 8 years, 3 months ± 1 year, 6 months) who underwent a cardiac procedure. The TOF group was compared with a group of children with acyanotic CHDs (ventricular septal defect [VSD], atrial septal defect, aortic stenosis or pulmonic stenosis) and with a healthy control group. In addition to age and sex, the groups were matched on the educational level of both parents and on the educational level of the child. Local school boards providing normal full-time education were contacted with specific demands to find a child matching the TOF child on sex, age, and educational level of the mother and father. All eligible children were tested at school. The local ethical committee approved the study, and all parents provided written informed consent. Procedures were in accordance with the recommendations found in the Helsinki Declaration of 1975.[11] Medical and surgical data were collected from the patients' files.

Intellectual and Neuropsychological Assessment

After parental agreement to participate, the child was invited for an intellectual and neuropsychological assessment of half a day.

The child was tested with a short form of the Wechsler Intelligence Scale for Children, 3rd edition, Dutch version. The short form included the subtests Information, Vocabulary, Picture Completion, and Block Design.[12] A deviation IQ was calculated by using the procedure suggested by Sattler.[13] On the subtest level, a mean performance of 10 (standard deviation [SD], 3) is expected. Mean estimated full-scale IQ is 100 (SD, 15).

The neuropsychological battery consisted of all core subtests of the NEPSY (a developmental neuropsychological assessment). The NEPSY tests the child's neuropsychological development in 5 functional domains to detect subtle deficiencies within and across these functional domains, which can interfere with learning in preschool- and school-aged children.[14] The 5 domains are Attention and Executive Functioning, Memory, Language, Visuospatial Skills, and Sensorimotor Functioning. A mean performance on the domains is 100 (SD, 15).

Behavioral Assessment

During the assessment of the child, the parents completed a behavioral questionnaire. The Child Behavior Checklist (CBCL) reports on the presence of behavioral, social, and emotional problems in 4- to 18-year old children, as reported by their parents. The CBCL contains both competence scales and problem behavior scales. In the competence scales 27 items ascertain the child's activities, social involvement, and school performance. A Composite scale summarizes the total competence of the child. A *t* score of less than 37 reflects maladjusted behavior. In the behavior problem scales 113 questions have to be rated by the parents on a 3-point Likert scale to indicate the frequency of the behavior. The items cluster into 7 subscales: Withdrawn Behavior, Physical