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 $\pm$ 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

Copyright © 2007 by The American Association for Thoracic Surgery

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 $\pm$ 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

Table 1. Demographics and medical variables of the 3 groups

Group	TOF group (n = 18)	Acyanotic group (n = 17)	Control group (n = 18)	P value*
Age	8 y, 3 mo ± 1 y, 6 mo	9 y, 2 mo ± 1 y, 6 mo	8 y, 4 mo ± 1 y, 6 mo	.470
Sex	10 male, 8 female	7 male, 10 female	10 male, 8 female	.827
Birth weight (g)	2853 ± 883	3215 ± 637	3419 ± 511	.026†
Birth length (cm)	47.9 ± 3.7	49.2 ± 2.8	50.8 ± 1.8	.014†
Apgar score immediately after birth	<4: 33.3% 4-7: 0% 7-10: 66.7%	<4: 0% 4-7: 0% 7-10: 100%	<4: 0% 4-7: 0% 7-10: 100%	<.001‡
Apgar score after 10 min	<4: 22.2% 4-7: 0% 7-10: 77.8%	<4: 0% 4-7: 0% 7-10: 100%	<4: 0% 4-7: 0% 7-10: 100%	<.001‡
Symptoms	None: 13.3% Cyanosis: 53.3% Heart murmur: 13.4% ≥2 of these symptoms: 20%	Shortness of breath: 22.2% Feeding difficulties: 22.2% Heart murmur: 11.1% Upper airway infections: 22.2% Excessive sweating: 11.1% ≥2 of these symptoms: 11.1%	—	—
Age at first operation	Median, 173 d	Median, 603 d	—	.014†
Duration of ECC (min)	89.2 ± 31.9	61.0 ± 33.0	—	.018†
Duration of aortic crossclamp (min)	51.6 ± 20.7	29.0 ± 21.3	—	.004†
Hypothermia (°C)	25	28	—	.010†

TOF, Tetralogy of Fallot; ECC, extracorporeal circulation. *Kruskal–Wallis test and Wilcoxon signed–rank test for 3 and 2 groups, respectively. Level of significance: † $P < .05$, ‡ $P < .001$.

Complaints, Anxious/Depressed Behavior, Social Problems, Thought Problems, Attention Problems, Delinquent Behavior, and Aggressive Behavior. The first 3 scales are grouped into a first global scale, Internalizing Behavior. The last 2 form a second global scale, Externalizing Behavior. All items grouped together constitute the Total Problem Behavior Scale. For each child, t scores ($M = 50$, $SD = 10$) were calculated. On the subscales, a t score of greater than 70 and on the global scales (Internalizing, Externalizing, and Total Problem Behavior) a t score of greater than 63 are considered to be indicative for clinical follow-up or treatment.^{15,16}

Statistical Analysis

Statistical analyses were performed with the SPSS for Windows statistical software package (version 12.0; SPSS, Inc). Demographics (age at the moment of testing, sex, and educational level of both parents), medical characteristics (birth weight, Apgar scores, duration of extracorporeal circulation, duration of aortic cross-clamp, and hypothermia), and outcome measures (IQ, NEPSY, and CBCL) were compared between the patient groups and control group by means of χ^2 statistics, Wilcoxon signed-rank tests, or Kruskal–Wallis tests. The term *significant* was used to indicate purely statistical and not clinical significance.

Results

Patient Characteristics and Medical Data

Comparison of the 3 groups (the TOF, acyanotic CHD, and healthy control groups) shows the lowest weight ($P < .05$) and length ($P < .05$) at birth in the TOF group, and also a larger part of this group has lower Apgar

scores immediately ($P < .001$) and 10 minutes ($P < .001$) after birth. Not surprisingly, in the TOF group the most frequent symptom was cyanosis, whereas in the acyanotic group presenting symptoms were various. The larger part of our TOF group had 1 operation (77.8%). The maximum number of operations a child underwent was 3. Seven patients had initial shunts. Median age at the time of the first operation was significantly lower in the TOF group than in the acyanotic CHD group ($P < .05$). All patients were operated on using standard procedures for extracorporeal circulation during moderate hypothermia (25°C–28°C). Circulatory arrest was not used. The TOF group underwent significantly longer periods of extracorporeal circulation and aortic cross-clamping ($P < .01$). At the moment of testing, all children were considered hemodynamically stable and in good health. Results can be found in Table 1.

Intellectual and Neuropsychological Assessment

We could not identify any significant differences between the TOF group and the acyanotic CHD group. Children in the TOF group showed significantly lower scores on the estimated Full Scale IQ ($P < .05$) than their healthy control subjects. On the NEPSY domains Language ($P < .01$) and Sensorimotor Functioning ($P < .01$) and on the subtests Tower ($P < .05$), Memory for Names ($P < .05$), Narrative Memory ($P < .05$), and Design Copy ($P < .05$)

Table 2. Group differences on intellectual and neuropsychological performances

Variable (mean $\pm$ SD)	TOF group (n = 18)	Acyanotic group (n = 17)	Control group (n = 18)	P value*
Intellectual performances (IQ: 100 $\pm$ 15, subtest: 10 $\pm$ 3)				
Estimated Full Scale IQ	94.6 $\pm$ 14.1	97.5 $\pm$ 16.3	106.2 $\pm$ 15	.014†
Picture Completion	8.4 $\pm$ 3.1	8.1 $\pm$ 4.1	10.0 $\pm$ 4.2	.033†
Block Design	9.0 $\pm$ 3.0	9.9 $\pm$ 3.1	10.3 $\pm$ 2.2	.229
Information	10.0 $\pm$ 2.9	10.8 $\pm$ 2.9	11.6 $\pm$ 2.0	.076
Vocabulary	9.7 $\pm$ 3.0	9.8 $\pm$ 2.0	11.0 $\pm$ 2.7	.016†
Neuropsychological performances (domains: 100 $\pm$ 15, subtests: 10 $\pm$ 3)				
Attention and Executive Functioning	112.0 $\pm$ 13.5	112.9 $\pm$ 7.1	117.0 $\pm$ 8.5	.070
Tower	12.6 $\pm$ 2.6	13.1 $\pm$ 1.9	14.1 $\pm$ 2.0	.036†
Auditory Attention/Response set	12.1 $\pm$ 1.3	12.6 $\pm$ 1.0	12.1 $\pm$ 1.3	.176
Visual Attention	10.0 $\pm$ 3.3	9.3 $\pm$ 2.5	10.4 $\pm$ 2.1	.092
Memory	99.4 $\pm$ 13.1	99.3 $\pm$ 17.0	103.0 $\pm$ 9.5	.232
Memory for Faces	11.0 $\pm$ 3.3	9.8 $\pm$ 3.6	9.6 $\pm$ 2.5	.277
Memory for Names	9.6 $\pm$ 3.0	8.4 $\pm$ 4.0	11.2 $\pm$ 1.8	.022†
Narrative Memory	9.1 $\pm$ 2.5	11.9 $\pm$ 2.9	10.4 $\pm$ 1.6	.030†
Language	101.1 $\pm$ 12.8	101.9 $\pm$ 12.3	115.7 $\pm$ 14.8	<.001‡
Phonological Processing	11.1 $\pm$ 3.1	11.3 $\pm$ 3.3	13.4 $\pm$ 3.2	.003†
Speeded Naming	9.1 $\pm$ 2.3	9.2 $\pm$ 2.5	10.9 $\pm$ 2.1	.011†
Comprehension of Instructions	10.6 $\pm$ 2.1	10.6 $\pm$ 2.2	12.7 $\pm$ 2.8	<.001‡
Visuospatial Skills	110.6 $\pm$ 17.3	117 $\pm$ 11.0	124.2 $\pm$ 11.6	.058
Design Copy	13.0 $\pm$ 3.3	14.9 $\pm$ 2.1	15.7 $\pm$ 1.8	.031†
Arrows	11.0 $\pm$ 3.5	10.7 $\pm$ 2.2	12.0 $\pm$ 2.4	.141
Sensorimotor Functioning	90.9 $\pm$ 10.6	86.7 $\pm$ 15.2	100.9 $\pm$ 10.7	<.001‡
Finger Tapping	9.1 $\pm$ 1.3	9.6 $\pm$ 1.4	10.4 $\pm$ 1.7	.021†
Imitating Hand Positions	9.3 $\pm$ 2.8	7.7 $\pm$ 2.8	11.8 $\pm$ 2.2	<.001‡
Visuomotor Precision	8.0 $\pm$ 2.6	7.3 $\pm$ 3.5	9.1 $\pm$ 2.7	.022†

SD, Standard deviation; TOF, tetralogy of Fallot. *Kruskal Wallis test. Level of significance: † $P < .05$, ‡ $P < .01$.

group differences became apparent. The small sample of children undergoing multiple operations does not permit statistical analysis, but qualitative analysis of the results does not show obvious differences between the children with a single operation for TOF or multiple operations. Mean performances on IQ and NEPSY can be found in [Table 2](#).

Behavioral Assessment

We could not identify any significant differences between the TOF group and the acyanotic CHD group. Parental reports revealed significantly higher scores on Attention Problems ($P < .05$) and the Total Problem Scale ($P < .05$) in the TOF group compared with in the healthy control subjects. The parents of the children with TOF also rated their child's school performances to be significantly lower than these of healthy peers ($P < .01$). The small sample of children undergoing multiple operations does not permit statistical analysis, but qualitative analysis of the results does not show obvious differences between the children with a single operation for TOF or multiple operations. Results on the CBCL can be found in [Table 3](#).

Discussion

In this study we defined the intellectual capacities, the neuropsychological performances, and the behavioral functioning of children with TOF 6 to 12 years postoperatively.

We compared the performances of the children in the TOF group with those of children in the acyanotic CHD group and those of healthy children. We could not identify differences in functional outcome between the TOF group and the acyanotic CHD group. This finding is in accordance with previous studies reporting similar intellectual⁵ and neuropsychological functioning in children with cyanotic (TOF and TGA) and acyanotic (VSD) CHDs.⁴

Compared with a group of healthy peers, the children in the TOF group showed lower intellectual abilities, specifically on the subtests Picture Completion and Vocabulary. However, only 3 children had an IQ of less than 85, confirming that although the intellectual capacities are lower than in the healthy group, the larger part of the children with TOF have a normal level of intelligence.^{5,6}

Concerning the neuropsychological profile, we found significant differences between the TOF group and the healthy group on language and sensorimotor functioning.

Table 3. Group differences on behavioral functioning (the Child Behavior Checklist)

Variable	TOF group (n = 18)	Acyanotic group (n = 17)	Control group (n = 18)	P value*
Competence scales (mean $\pm$ SD; <i>t</i> score <37: clinically significant score)				
Activity scale	47.7 $\pm$ 6.6	46.4 $\pm$ 9.1	49.0 $\pm$ 4.7	.526
Social scale	48.6 $\pm$ 8.1	47.3 $\pm$ 8.2	47.7 $\pm$ 7.3	.515
School scale	40.2 $\pm$ 7.1	45.2 $\pm$ 8.7	51.1 $\pm$ 7.3	<.001†
School results	Insufficient: 5.6% Weak: 16.7% Average: 61.1% Good: 16.7%	Insufficient: 0% Weak: 12.5% Average: 31.3% Good: 56.3%	Insufficient: 0% Weak: 0% Average: 27.8% Good: 72.2%	.001†
Special education	Yes: 5.6% No: 94.4%	Yes: 0% No: 100%	Yes: 0% No: 100%	.268
Repeating a school year	Yes: 16.7% No: 83.3%	Yes: 18.8% No: 81.3%	Yes: 5.6% No: 94.4%	.022†
School problems	Yes: 66.7% No: 33.3%	Yes: 56.3% No: 43.8%	Yes: 22.2% No: 77.8%	<.001†
Problem behavior scales (mean $\pm$ SD, <i>t</i> score >63; for individual scales, >70: clinically significant score)				
Internalizing Behavior	51.1 $\pm$ 11.1	53.0 $\pm$ 10.6	48.4 $\pm$ 7.1	.051
Withdrawn	54.9 $\pm$ 7.5	54.1 $\pm$ 5.9	52.1 $\pm$ 3.5	.065
Physical Complaints	56.8 $\pm$ 9.8	57.1 $\pm$ 7.4	51.9 $\pm$ 3.1	.109
Anxious/Depressed	53.7 $\pm$ 6.4	55.4 $\pm$ 6.8	52.7 $\pm$ 4.3	.479
Externalizing Behavior	52.8 $\pm$ 10.7	52.6 $\pm$ 11.9	49.4 $\pm$ 7.6	.137
Delinquent Behavior	53.5 $\pm$ 5.4	54.8 $\pm$ 6.8	50.7 $\pm$ 2.3	.173
Aggressive Behavior	56.4 $\pm$ 7.4	57.1 $\pm$ 9.5	53.4 $\pm$ 4.3	.186
Social Problems	53.5 $\pm$ 4.1	54.8 $\pm$ 6.2	51.3 $\pm$ 2.4	.051
Thought Problems	54.9 $\pm$ 8.0	53.67 $\pm$ 4.7	53.5 $\pm$ 6.0	.099
Attention Problems	61.5 $\pm$ 9.1	59.9 $\pm$ 10.9	53.6 $\pm$ 5.1	.003†
Total Problem Behavior	56.0 $\pm$ 11.7	52.7 $\pm$ 14.2	47.6 $\pm$ 8.1	.003†

TOF, Tetralogy of Fallot; SD, standard deviation. *Kruskal-Wallis test. Level of significance: †*P* < .05, ‡*P* < .01.

Language deficits have been identified before in children with cyanotic CHDs,^{10,17} and in the TOF group higher rates of dysfunction in expressive and receptive language have been diagnosed compared with normative data.⁶ In our study the children with TOF had more difficulties, particularly with phonological awareness; had slower access to names of color, size, and shape; and lacked the ability to process and respond quickly to verbal instructions of increasing difficulty. These objective measures might be the cause of previously reported lower academic skills, such as reading and spelling, in children with CHDs.^{1,3}

We also found a group difference on sensorimotor functioning. The children with TOF performed worse than their healthy peers on subtests measuring motor speed and the ability to imitate finger and hand positions. In a comparative study on children with an acyanotic form of CHD (VSD) and children with TOF, the latter displayed a higher incidence of gross motor dysfunction.⁶ In our study, however, both the children with TOF and the children with acyanotic CHDs specifically showed slowness in simple and complex motor movements and an inefficient processing of tactile and kinesthetic information when they had to imitate finger and hand positions.

Performances on the subtest Tower were lower for the TOF group, indicating more difficulties in tasks concerning executive functioning, which was also reported in a study on adult patients with TOF.⁷ The hypothesis of damage in the prefrontal cortex is to be further examined.

Although obvious memory problems were not present, children with TOF did have lower scores than their healthy peers on a task measuring narrative memory. Possibly it was not the memory aspect that caused this lower score but rather the lack of narrative skills. In a study on narrative discourse in children with a cyanotic form of CHD (TGA) 4 years after surgical intervention, their performance differed in frequency and diversity of narrative elements and in adequacy of the information provided.¹⁸ Because the number of narrative elements the child can reproduce constitutes the narrative memory score in the present study, this seems a plausible explanation for the lower scores in our TOF children. Furthermore, memory for names also appeared worse in children with TOF than in healthy control subjects. The poor performance might be related to difficulties in recalling names and a deficient retrieval of sound-symbol association and spoken-written word connections, which can be reduced to deficient linguistic skills.

Visuospatial skills constituted another domain in which children with TOF performed worse than healthy control subjects. Their ability to copy 2-dimensional geometric figures was reduced. We hypothesize that the lower score on design copy was caused by hand-eye coordination difficulties and visual perceptual deficits, which have been previously reported in children after infant cardiac surgery.^{1,17,19}

The behavioral functioning of children with TOF in our study group, measured by means of a parental questionnaire, is characterized by mainly attention and school problems. These results contradict the previously reported absence of behavioral problems in children with cyanotic CHDs.^{1,10} Other studies, however, did report a higher incidence of behavioral problems,⁹ specifically a higher risk for attention problems.²⁰ These attention problems might have led, at least partially, to the significantly lower school competency in TOF children reported by the parents. Specifically, lower school results and more school problems were indicated. This finding is in accordance with follow-up studies in adolescents and adults with CHDs that report patients to have spent longer periods in school,²¹ display more need for special education, and more often present with a learning disability.²² Neuropsychological deficits that have been mentioned before in outcome studies^{6,10,17} are probably responsible for the shortcomings at the school level.

The small sample size constitutes a limitation in our study. Furthermore, we cannot ignore a possible selection bias in the patient groups. Parents of children with a CHD voluntarily entered the study. The suspicion of a neurodevelopmental or behavioral problem in their child might have been the parent's motivation for participation, whereas parents of children who perform well at school might have wished not to stigmatize their child as a patient. In accordance with previous studies,²³ our data reflected lower birth weight in the TOF group. However, this birth weight is still within the normal range (2853 g). Adverse cognitive outcome has been described in children with very low (<1501 g) and extremely low (≤ 1000 g) birth weight.²⁴ For this reason, we believe the relatively lower birth weight in the TOF group was of little influence on the reported neuropsychological outcome.

In this outcome study we compared the intellectual, neuropsychological, and behavioral function of children with TOF 6 to 12 years postoperatively with that of a group with acyanotic CHDs and healthy peers. We could not identify any functional outcome differences between the TOF group and the acyanotic group. In the TOF group we identified a lower estimated full-scale intelligence and a neuropsychological profile characterized by mainly mild motor deficits and difficulties with language tasks. Attention problems and lower school competencies are the most important behavioral aspects reported by parents of children

with TOF. In general, our findings are comparable with those of most outcome studies in children with CHD. However, our study resulted in the identification of specific neuropsychological shortcomings in children with CHD, which can lead to tailored interventional programs focused especially on motor functions and language. By means of these programs, the mentioned school problems and the long-term neuropsychological sequelae might be prevented.

References

1. Wright M, Nolan T. Impact of cyanotic heart disease on school performance. *Arch Dis Child.* 1994;71:64-70.
2. Wray J, Sensky T. How does the intervention of cardiac surgery affect the self-perception of children with congenital heart disease? *Child Care Health Dev.* 1998;24:57-72.
3. Wray J, Sensky T. Congenital heart disease and cardiac surgery in childhood: effects on cognitive function and academic ability. *Heart.* 2001;85:687-91.
4. Oates R, Simpson J, Cartmill T, Turnbull J. Intellectual function and age of repair in cyanotic congenital heart disease. *Arch Dis Child.* 1995;72:298-301.
5. Haneda K, Itoh T, Togo T, Ohmi M, Mohri H. Effects of cardiac surgery on intellectual function in infants and children. *Cardiovasc Surg.* 1996;4:303-7.
6. Hövels-Gürich H, Konrad K, Skorzenski D, Nacken C, Minkenberg R, Phys D, et al. Long-term neurodevelopmental outcome and exercise capacity after corrective surgery for tetralogy of Fallot or ventricular septal defect in infancy. *Ann Thorac Surg.* 2006;81:958-67.
7. Daliento L, Mapelli D, Russo G, Scarso P, Limongi F, Iannizzi P, et al. Health related quality of life in adults with repaired tetralogy of Fallot: psychosocial and cognitive outcomes. *Heart.* 2005;91:213-8.
8. Utens E, Verhulst FC, Meijboom FJ, Duivenvoorden HJ, Erdman RA, Bos E, et al. Behavioural and emotional problems in children and adolescents with congenital heart disease. *Psychol Med.* 1993;23:415-24.
9. Oates RK, Turnbull JA, Simpson JM, Cartmill TB. Parent and teacher perceptions of child behaviour following cardiac surgery. *Acta Paediatr.* 1994;83:1303-7.
10. Bellinger DC, Rappaport LA, Wypij D, Wernovsky G, Newburger JW. Patterns of developmental dysfunction after surgery during infancy to correct transposition of the great arteries. *J Dev Behav Pediatr.* 1997;18:75-83.
11. World Medical Association Declaration of Helsinki. Recommendations guiding physicians in biomedical research involving human subjects. *JAMA.* 1997;277:925-6.
12. Grégoire J. Comparison of three short forms of the Wechsler Intelligence Scale for children—third edition (WISC-III). *Eur Rev Appl Psychol.* 2000;50:437-41.
13. Sattler J. Assessment of children. 3rd ed. San Diego, Calif: Author; 1992.
14. Korkman M, Kirk U, Kemp S. NEPSY. A developmental neuropsychological assessment. San Antonio, Texas: The Psychological Corporation. A Harcourt Assessment Company; 1998.
15. Achenbach TM. Manual for the child behaviour checklist/4-18 and 1991 profile. Burlington, Vt: University of Vermont; 1991.
16. Verhulst FC, van der Ende J, Koot HM. Handleiding voor de CBCL/4-18 [Manual of the CBCL/4-18]. Rotterdam, The Netherlands: Universiteit Rotterdam; 1996.
17. Hövels-Gürich HH, Seghaye MC, Schnitker R, Wiesner M, Huber W, Minkenberg R, et al. Long-term neurodevelopmental outcomes in school-aged children after neonatal arterial switch operation. *J Thorac Cardiovasc Surg.* 2002;124:448-58.
18. Hemphill L, Uccelli P, Winner K, Chang CJ, Bellinger D. Narrative

- discourse in young children with histories of early corrective heart surgery. *J Speech Lang Hear Res.* 2002;45:318-31.
19. Bellinger D, Bernstein J, Kirkwood M, Rappaport L, Newburger J. Visual-spatial skills in children after open-heart surgery. *J Dev Behav Pediatr.* 2003;24:169-79.
20. O'Dougherty M, Berntson G, Boysen S, Wright F, Teske D. Psychophysiological predictors of attentional dysfunction in children with congenital heart defects. *Psychophysiology.* 1988;25:305-15.
21. Ternstedt BM, Wall K, Oddsson H, Riesenfeld T, Groth I, Schollin J. Quality of life 20 and 30 years after surgery in patients operated on for Tetralogy of Fallot and for atrial septal defect. *Pediatr Cardiol.* 2001; 22:128-32.
22. Nolan T, Pless B. Emotional correlates and consequences of birth defects. *J Pediatr.* 1986;109:201-16.
23. Kramer HH, Trampisch HJ, Rammos S, Giese A. Birthweight of children with congenital heart disease. *Eur J Paediatr.* 1990;149:752-7.
24. Böhm B, Katz-Salamon M, Smedler AC, Lagercrantz H, Forssberg H. Developmental risks and protective factors influencing cognitive outcome at 5½ years of age in very-low-birthweight children. *Dev Med Child Neurol.* 2002;44:508-16.