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Long-term outcome of speech and language in children after corrective surgery for cyanotic or acyanotic cardiac defects in infancy

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ABSTRACT

The purpose of this prospective study was to assess whether outcome of speech and language in children 5-10 years after corrective surgery for tetralogy of Fallot (TOF) or ventricular septal defect (VSD) in infancy was influenced by the preoperative condition of hypoxemia or cardiac insufficiency and whether it was associated with perioperative risk factors and neurodevelopmental outcome.

A total of 35 unselected children, 19 with TOF and hypoxemia and 16 with VSD and cardiac insufficiency, operated with combined deep hypothermic circulatory arrest and low-flow cardiopulmonary bypass at mean age 0.7 ± 0.3 (mean ± standard deviation) years, underwent, at mean age 7.4 ± 1.6 years, standardized evaluation of speech and language functions. Results were compared between subgroups and related to perioperative factors, sociodemographic and neurodevelopmental status.

Age at testing, socioeconomic status and history of speech and language development were not different between the subgroups. In contrast, total scores on oral and speech motor control functions (TFS) as well as on oral and speech apraxia (Mayo Test) were significantly reduced (p<0.02 to <0.05), and scores on anatomical oral structures tended to be lower (p<0.09) in the TOF group as compared to the VSD group. No differences were found for auditory word recognition and phonological awareness as assessed by the Auditory Closure subtest of the Illinois Test of Psycholinguistic Abilities and the test of auditory analysis skills, respectively. In all children, higher age at testing and better socioeconomic status were associated with better results in all domains of assessment (p<0.001 to <0.04). Consistent impairments of all oral and speech motor control functions (TFS and Mayo Test) were present in 29% of all children with a mean age of 6.5 years in contrast to 43% with normal performance and a mean age of 8.3 years. On the receptive speech tasks, only 6% scored below the normal range of their age group. TFS subscores were significantly correlated with age, bypass duration and motor function, but not correlated with socioeconomic status, duration of cardiac arrest, intelligence and academic achievement.

Children with preoperative hypoxemia due to cyanotic cardiac defects in infancy are at higher risk for dysfunction in speech and language than those with cardiac insufficiency due to acyanotic heart defects. Age at testing, socioeconomic status, and duration of cardiopulmonary bypass influenced test results. Long-term outcome in speech and language functions can be considered as a sensitive indicator of overall child development after cardiac surgery.

Keywords : Speech and language development ; Tetralogy of Fallot ; Ventricular septal defect ; Cardiac surgery

Abbreviations : AC, auditory closure subtest of ITPA ; CPB, cardiopulmonary bypass ; DHCA, deep hypothermic cardiocirculatory arrest ; FISH, fluorescence in situ hybridization ; IQ, intelligence quotient ; ITPA, Illinois test of psycholinguistic abilities ; K-ABC, Kaufman assessment battery for children ; MQ, motor quotient ; OFS, oral functional score ; SD, standard deviation ; SFS, speech functional score ; TAAS, test of auditory analysis skills ; TFS, total functional score ; TOF, tetralogy of Fallot ; TSS, total structural score ; VSD, ventricular septal defect.

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1. Introduction

Children after cardiac operations for congenital heart defects with cardiopulmonary bypass (CPB) with or without hypothermic cardiac arrest generally are at risk of later neurodevelopmental impairment in the domains of motor functions, intelligence, academic achievement, attention, speech, language and behavior.[1-15] In particular, we recently showed that corrective surgery in infancy for tetralogy of Fallot (TOF) or ventricular septal defect (VSD) with combined circulatory arrest and low flow bypass is associated with reduced neurodevelopmental outcome with respect to motor function, formal intelligence and academic achievement.[14] We additionally demonstrated that children with preoperative hypoxemia in infancy due to cyanotic heart defects are at higher risk for motor dysfunction[14] and attentional dysfunction in the field of executive control[15] than are children with cardiac insufficiency due to acyanotic heart defects.

As a milestone of child development, speech and language functions are of special interest with respect to the long-term follow-up of children after cardiac surgery. However, detailed systematic studies are rare in this field.[2,3,11,16] Given the high prevalence of speech and language dysfunctions in association with motor, cognitive and academic disabilities, speech and language dysfunctions are considered to be an important indicator of general developmental impairment.[2,17-19]

The present prospective study was designed to test the hyposthesis that (1) the preoperative condition of hypoxemia in infancy is associated with worse outcomes in speech and language later in childhood, when compared to the condition of preoperative cardiac insufficiency; furthermore, (2) the relationship between speech and language outcomes and perioperative management, neurodevelopmental outcomes and sociodemographic status should be evaluated in a uniform cohort of children with TOF or VSD at age 5-11 years after corrective surgery in infancy.

2. Methods

2.1. Patient population

Between 1993 and 1999, 24 consecutive infants with a regular form of TOF and hypoxemia and 26 consecutive infants with a VSD and cardiac insufficiency had undergone primary corrective cardiac surgery in our institution at a mean age of 0.7 years with a standard deviation (SD) of 0.3. There was no early or late mortality. All patients with TOF had normal chromosomal status (46, XY and XX, respectively), and microdeletion 22q11.2 had been excluded by fluorescence in situ hybridization (FISH) analysis. In 2004, at a mean age of 7.4 years (SD 1.6, range 5.0-11.8), we performed a follow-up study of neurodevelopmental outcome and exercise capacity (14), attentional functions (15) as well as outcome of language and speech development.

Ten of the original 50 patients could not present for follow-up examination (4 with TOF and 6 with VSD). They were considered by a questionnaire completed by the parents as well as by the treating pediatricians and pediatric cardiologists. Actual participation in the study was mainly determined by the distance of the family's residence from our institution. The children who did not participate were similar to participants with respect to age, sex, cardiac health, physical activity, neurodevelopmental outcome and socioeconomic background.

The primary study group coming to the reevaluation consisted of 40 unselected children (80% of the operated patients). Among these, the results of speech and language examination of 5 patients could not be considered, as they were incomplete because of the children's Dutch mother tongue (3 patients with VSD), insufficient compliance (1 patient with VSD) or lacking time management of the family (1 patient with TOF).

The final study group for the assessment of language and speech therefore consisted of 35 patients (70% of the operated patients) with a mean age of 7.6 years (SD 1.6). Subgroup A consisted of 19 patients with TOF (53% male, age at surgery 0.7 ± 0.3 years, age at evaluation 7.5 ± 1.4 years). All group A patients had suffered from preoperative cyanosis (arterial oxygen saturation <90%). Group B consisted of 16 patients with VSD (50% male, age at surgery 0.7 ± 0.2 years, age at evaluation 7.8 ± 1.9 years). All group B patients had suffered from clinical signs of heart failure and pulmonary hypertension. At the time of evaluation, all patients were in a good general and cardiac health status, and none of them received any cardiac-related medication. All patients had anatomically and functionally adequate repair of their defects. Socioeconomic status of the total group as well as of the subgroups[20] was not different from that of a normal population.[14] Written informed consent of the parents was obtained.

2.2. Surgical management and perfusion methods

Corrective surgery was performed following standardized deep hypothermic circulatory arrest (DHCA, 52.9 ± 8.8 min) with a minimal nasopharyngeal temperature of 15.2 ± 1.9 °C and low-flow CPB (60.7 ± 20.3min). The protocols of the bypass technique and of the postoperative care have been described in detail previously.[14]

2.3. Perioperative and neurodevelopmental risk factors

The patients' data with respect to perioperative management as well as preoperative, perioperative, and postoperative complications were prospectively registered at time of surgery and have been recently published.[14] The neurodevelopmental follow-up examination[14] consisted of a standardized test battery evaluating a broad spectrum of motor and cognitive development.

As shown in Table 1, the children with TOF and VSD (subgroup A and B) did not differ with respect to age at testing, socioeconomic status, and durations of DHCA and CPB. However, the two subgroups differed significantly in gross motor functions. Children with TOF as a group showed a motor quotient of 79.9 which is below the normal range of 85-115. In contrast, no significant differences were present with respect to general intelligence and acquired abilities. Both groups showed quotients between 90 and 95. The present study started from the question of how these characteristics were related to outcome parameters of language and speech. Would the TOF group also show a less favorable development of speech and language?

Table 1 *- Sample characteristics at follow-up examination*[14]

	Total group *n* = 35	Subgroup A TOF *n* = 19	Subgroup B VSD *n* = 16	*P* value[a] A vs. B
Age at testing (years)	7.6 ± 1.6	7.5 ± 1.4	7.8 ± 1.9	0.66
Socioeconomic status[b]	2.2 ± 1.1	2.1 ± 1.2	2.4 ± 0.9	0.35
Duration of DHCA (min)	52.9 ± 8.8	54.7 ± 6.9	50.6 ± 10.6	0.26
Duration of CPB (min)	60.7 ± 20.3	63.8 ± 20.6	57.1 ± 20.0	0.27
Motor quotient (MQ)[c]	85.7 ± 13.3	79.9 ± 9.5	92.2 ± 14.1	0.01
Intelligence quotient (IQ)[d]	91.9 ± 13.1	92.0 ± 13.3	91.8 ± 13.3	0.91
Acquired abilities[e]	94.0 ± 14.6	94.1 ± 15.6	93.9 ± 13.8	0.95

DHCA, deep hypothermic circulatory arrest; CPB, cardiopulmonary bypass.TOF, tetralogy of Fallot; VSD, ventricular septal defect.Values are mean±SD, standard deviation.
[a] P values calculated by Mann-Whitney *U*-test for comparison of central tendencies between subgroups.
[b] Social classification of the family according to Habich and Noll.[20]
[c] Standard score of gross motor function of the Kiphard and Schilling body coordination test.[21]
[d] Standard score of general intelligence of the Kaufman Assessment Battery for children.[22]
[e] Standard score of acquired abilities (learning, knowledge) of the Kaufman Assessment Battery for children.[22]

2.4. Outcome parameters

2.4.1. Speech and language history

A short interview with a duration of about 10 min was conducted by a licensed Speech and Language Therapist (coauthor SBB) to obtain from the parents (mother, father, or both of them) data on speech and language development, hearing impairment, family disposition for speech and language disorders, social circumstances and speech therapy.

The parents were asked about present and past risks and developmental disorders.

2.4.2. Examination of speech and language

The examination was performed by the speech and language therapist in a uniform manner during the early afternoon and lasted about 30 min. At the time of assessment, all children had normal faculty of hearing as assessed by pure tone threshold audiometry. Language and speech was examined by means of 4 standardized tests that were already used by Bellinger and collegues[10] in the Boston Circulatory Arrest Trial and in our previous study on patients after arterial switch operation.[2] They were adapted to German. All responses were

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online protocolized and/or audio recorded for later detailed evaluation.

The Oral and Speech Motor Control Protocol,[23] adapted to German by coauthors RSch and WH is targeting at 7 functional components of the vocal tract and their corresponding cranial nerves: lips, lower jaw (mandible), upper jaw (maxilla), teeth, tongue, velopharynx and larynx-respiration. In addition, coordination of speech movements is assessed by single word repetitions, and a speech sample (5-min of free conversation) is elicited for a scoring of speech rate, melody and voice. In total, the protocol comprises 80 items. Performance on each item is scored as normal (1 or 2 points), subnormal (1 point) or not normal (0 points). A 'total structural score' (TSS) with a maximum of 24 points is calculated from the structures at rest. A 'total functional score' (TFS) with a maximum of 112 points is calculated from the functional structures, subdivided into an 'oral functional score' (OFS) with a maximum of 32 points and a 'speech functional score' (SFS) with a maximum of 80 points. Furthermore, maximum speech performance measures are examined: phonation time of the vowel /a:/ (normal value > 10.0 s for children 6-12 years of age), monosyllabic repetition rate (normal value > 4.0 syllables per second for children 6-12 years of age), and polysyllablic repetition rate (normal value >5.0 syllables per second for children 6-12 years of age).

The Mayo Test of Speech and Oral Apraxia (Children's Battery),[24] adapted to German by coauthors RSch and WH, serves to assess motor speech functions as well as orofacial non-verbal movements. The German version contains 26 selected items of the battery, divided into three parts. Performance on each item is scored from correct (8 points) to impossible (0 points). Part A with 20 items and a maximum of 160 points contains verbal and—on failure-nonverbal instructions to perform orofacial movements, part B with two items and a maximum of 16 points requires imitation of multiple orofacial movements, part C with four items and a maximum of 32 points demands repetition of sentences. In summary, a total maximum of 208 points can be achieved.

The Auditory Closure (AC) subtest of the Illinois Test of Psycholinguistic Abilities (ITPA),[25] German version by Anger-maier,[25] aims at phonological decoding, lexical recognition and production of single words in children aged 3-10 years. Stimulus words spoken by the examiner with missing sounds have to be completed and repeated correctly. The total number of 36 items of the original test was reduced to 30 for the purpose of this study (cf. Bellinger et al.[10]). The total score consists of a maximum of 30 points, one for each correctly recognized and spoken item.

The Test of Auditory Analysis Skills (TAAS),[26] adapted to German by coauthors RSch and WH, also requires auditory comprehension in preschool and school age children. In particular, capabilities of phonological awareness and processing are investigated. Verbally presented single words must be first repeated completely and then repeated again with omission of specific morphemes or phonemes as instructed by the examiner. The maximum score consists of 13 points, one for each correctly spoken item.

In order to assess the relationship between the different tests described above, patients were classified as having a normal or an impaired overall speech function by means of the total functional score (TFS) with respect to function of volitional oral movements, articulation, phonation and coordination of speech. The cut-off point for an impaired speech function was set at a score of less than 104 points which is equivalent to the minimum performance of the validation sample.[23]

2.5. Statistical analysis

Results were expressed by mean values and standard deviation (m ± SD), or as percentages. For comparison of frequencies by means of contingency tables, Fisher's exact test was used. For intergroup comparison, the non-parametric Mann-Whitney *U*-test was administered. The Spearman rank correlation coefficient was assessed for correlation of independent parameters. Alpha-adjustment for repeated measures was done according to Bonferroni-Holm.

Statistical analysis was performed with the SPSS for Windows software, version 14.0 (SPSS GmbH Software, München, Germany). All statistical tests were performed at a significance level of 0.05. The term "significant" was only used to indicate statistical, not clinical significance.

***Table 2** - Parental reports on speech and language history: Number and percentage of children with present or past risk and disorder*

	Total group	Subgroup A TOF	Subgroup B VSD	P value[a]

	n = 35		*n* = 19		*n* = 16		A vs. B
Familial deficits of language and speech	13	37.1	8	42.1	5	31.3	0.38
Siblings with speech/language disorders	4	16.0	2	14.3	2	18.2	0.60
Bilingual language development	7	20.0	3	15.8	4	25.0	0.40
Delayed onset of language development	11	31.4	7	36.8	4	25.0	0.35
Impaired development of language and/or speech	29	82.9	16	84.2	13	81.3	0.58
Stuttering	5	14.3	1	5.3	4	25.0	0.12
Cluttering	12	34.3	5	26.3	7	43.8	0.23
Voice disorder	3	8.6	1	5.3	2	12.5	0.43
Articulation disorder (Dyslalia)	23	65.7	14	73.7	9	56.3	0.23
Delayed lexical development	4	11.4	3	15.8	1	6.3	0.37
Delayed syntactic development	8	22.9	4	21.1	4	25.0	0.55
Dysgrammatism	7	20.0	3	15.8	4	25.0	0.40
Dyslexia/dysgraphia (school children, *n* = 23)	9	39.1	5	41.7	4	36.4	0.57
Speech/language therapy	13	37.1	9	47.4	4	25.0	0.16
Other therapy (physio/occupational/play therapy)	9	25.7	8	42.1	1	6.3	0.02

TOF, tetralogy of Fallot; VSD, ventricular septal defect.
[a] *P* values calculated by Fisher's exact test for comparison between subgroups (1-tailed).

3. Results

3.1. Parental report on speech and language history

Table 2 shows the results of the parents' interview with respect to familial disposition for speech problems and to several aspects of speech development of their children. The proportions of reported developmental risks and disorders were high in most instances and exceeded the typical prevalence rates of 2-10% that are estimated for the global population.[27] Almost half of the TOF children in contrast to 25% of the VSD children received speech/language therapy. However, none of the differences between subgroups were significant. In contrast, other therapy forms were significantly more frequently reported for TOF than for VSD children (see Table 2). Severe diseases of the throat, nose and ears or hearing deficiencies were not reported. All patients had undergone periodical assessment of hearing.

3.2. Outcomes for speech and language

The results are given in Table 3. On the Oral and Speech Motor Control Protocol, the total group had lower mean scores on both the TSS and the total functional score (TFS) than 4- to 7-year-old children of the US American validation sample.[23] The TOF group scored lower than the VSD group which was significant for the total functional score (TFS) and the entailed SFS. On maximum speech performance measures (vowel phonation time, monosyllabic and polysyllabic speech rate), the two subgroups did not differ, but the obtained mean values were below the normal range of children of the same age group. To estimate the degree of impairment, we compared the individual TSS and TFS scores to the minimum performance of the validation sample (18 and 104 points, resp.). On TFS scores, 15 children (11 of the TOF subgroup, 58%, and 4 of the VSD subgroup, 25%) scored below the normal range of 4- to 7-year-old children of the American validation sample, i.e. they must be considered to be impaired in their speech development. Note that such a cross-linguistic comparison was justified as the TFS focuses on general oral motor and speech functions and not on linguistic characteristics of the participants' mother tongue. In contrast, on TSS scores, only two children of the TOF subgroup (11%) scored below the normal range.

On the Mayo Test for Apraxia of Speech and Oral Apraxia, the subscore on multiple orofacial movements (part B) as well as the total score were also significantly reduced in the TOF group compared to the VSD group, whereas the subscore on single orofacial movements (part A) tended to be reduced in the TOF compared to the VSD group. No differences were found for repetition of sentences (part C). All mean values were well below ceiling level which stands for normal performance. When individual Mayo total scores were compared to a mastery criterion of $P = 0.90$ (binomial distribution, alpha = 5%), 15 children (10 of the TOF and 5 of the VSD subgroup) did not reach criterion. Yet, all 15 children reached a criterion of $P = 0.70$, i.e. they presented with a mild degree of developmental oral apraxia.

On the Auditory Closure subtest of ITPA, there was no significant difference between the subgroups (see Table

3). Nearly all children showed average or even above average performance for their age when compared to the German standardization sample (*T* = 40 or more).[25] One child showed a severe deficit (*T* = 0) and one scored just below average (*T* = 38). Both belonged to the TOF subgroup.

Likewise, the TAAS did not show significantly different results between the subgroups. When individual scores were compared to age- and grade-related cut-offs reported for the US American validation sample,[26] only one child (from the TOF subgroup) was two grade levels behind. Ten more children (6 from the Fallot subgroup and 4 from the VSD subgroup) were one grade level behind. Their auditory word analysis skills were reduced, but did not classify as impaired. It must be stressed, however, that these estimates are preliminary as long as the German TAAS version is not validated separately. Note that neither a recent standardization of the German version of the ITPA nor of the American TAAS is available. Therefore, we cannot exclude the possibility that the auditory performance of the children would have been reduced when compared to a matched control group or the more recent norms.

Table 3 *- Outcomes of speech and language*

	Range of points	Total group *n* = 35	Subgroup A TOF *n* = 19	Subgroup B VSD *n* = 16	*P* value[a] A vs. B
Oral and Speech Motor Control Protocol					
Total structural score (TSS)	0-24	19.9 ± 1.6	19.5 ± 1.8	20.3 ± 1.2	0.08
Total functional score (TFS)	0-112	101.9 ± 7.2	96.3 ± 8.6	104.7 ± 3.7	0.02
Oral functional score (OFS)	0-32	29.0 ± 1.8	28.6 ± 2.1	29.4 ± 1.4	0.09
Speech functional score (SFS)	0-80	72.9 ± 6.7	71.0 ± 7.8	75.3 ± 4.1	0.03
Maximum phonation time (/a:/ in seconds)		8.8 ± 3.4	9.2 ± 3.5	8.3 ± 3.2	0.16
Monosyllabic speech rate (syllables per second)		4.4 ± 0.6	4.3 ± 0.7	4.5 ± 0.5	0.19
Polysyllabic speech rate (syllables per second)		4.4 ± 0.8	4.2 ± 0.8	4.5 ± 0.8	0.15
Mayotest for Apraxia of Speech *and Oral Apraxia (total score)*	0-208	179.0 ± 16.3	174.4 ± 18.2	184.5 ± 12.1	0.04
Part A: single orofacial movements	0-160	137.5 ± 10.5	134.8 ± 11.7	140.8 ± 8.0	0.08
Part B: multiple orofacial movements	0-16	14.4 ± 2.2	13.7 ± 2.5	15.3 ± 1.4	0.02
Part C: repetition of sentences	0-32	27.0 ± 6.5	25.8 ± 7.7	28.4 ± 4.5	0.18
Auditory Closure subtest *of* ITPA	0-30	17.0 ± 4.2	16.2 ± 4.7	17.9 ± 3.6	0.11
Test *of* Auditory Analysis Skills (TAAS)	0-13	7.1 ± 3.3	6.8 ± 3.4	7.4 ± 3.2	0.26

TOF, tetralogy of Fallot; VSD, ventricular septal defect.
Values are mean±SD, standard deviation.
[a] *P* values calculated by Mann-Whitney *U*-test for comparison of central tendencies between subgroups (1-tailed).

3.3. Differences between children with normal versus impaired oral and speech motor functions

In Table 4, the total group is separated between normal and impaired performance on TFS scores. The cut-off was below 104 points. The proportion of TOF children is higher in the impaired group. No differences were found with respect to gender, socioeconomic status and duration of DHCA. The impaired children were significantly younger, and duration of CPB was significantly longer in the impaired group. With

respect to performance variables, significant differences were found for Mayo total score (*P* = 0.02), the TAAS (*P* = 0.008) and for the standard score of gross motor function (MQ[21]).

Among the 15 children impaired on TFS, there were 10 children (7 from the TOF and 3 from the VSD subgroup) who scored subnormal on Mayo total score as well. They counted for 29% of the whole sample. In contrast, normal performance on both measures was obtained in 43% (5 from the TOF and 10 from the VSD subgroup). In addition, maximum phonation time was reduced in the consistently impaired group (mean 5 s (range 3-14) in contrast to 10 s (range 5-16) in the normal group). The children with consistently impaired oral and speech motor control functions were younger (mean 6.6 years (5.0-7.8) in contrast to 8.3 years (5.4-11.0) in the normal group).

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Table 4 *- Differences between children with normal versus impaired oral motor functions*

	TFS normal (*n* = 20)	TFS impaired (*n* = 15)	*P* value
TOF/VSD	8/12	11/4	[a]0.05
Male/female	10/10	8/7	[a]0.56
Age at testing (years)	8.2 ± 1.7	6.9 ± 1.3	[a]0.02
Socioeconomic status	2.2 ± 1.0	2.3 ± 1.2	[a]0.59
Duration of DHCA (min)	52.7 ± 10.3	53.3 ± 6.5	[a]0.90
Duration of CPB (min)	54.3 ± 15.7	69.3 ± 23.1	[a]0.02
Mayotest for Apraxia of Speech and Oral Apraxia (total score)	185.3 ± 10.5	170.6 ± 19.1	[b]0.02
Auditory Closure subtest of ITPA	17.7 ± 3.0	16.0 ± 5.4	[b]0.43
Test of Auditory Analysis Skills (TAAS)	8.3 ± 2.7	5.5 ± 3.4	[b]0.01
Motor quotient (MQ)[C]	90.8 ± 13.9	79.1 ± 9.2	[b]0.02
Intelligence quotient (IQ)[d]	93.4 ± 11.5	89.8 ± 15.1	[b]0.36
Acquired abilities[e]	94.2 ± 14.3	93.7 ± 15.5	[b]0.92

TFS, total functional score of the Oral and Speech Motor Control protocol.
ITPA, Illinois Test of Psycholinguistic Abilities. TOF, tetralogy of Fallot; VSD, ventricular septal defect. DHCA, deep hypothermic circulatory arrest; CPB, cardiopulmonary bypass.
Values are mean±SD, standard deviation.
[a] P values calculated by Fisher's exact test for comparison between subgroups (1-tailed).
[b] P values calculated by Mann-Whitney *U*-test for comparison of central tendencies between subgroups.
[c] Standard score of gross motor function of the Kiphard and Schilling body coordination test.[21]
[d] Standard score of general intelligence of the Kaufman Assessment Battery for children.[22]
[e] Standard score of acquired abilities (learning, knowledge) of the Kaufman Assessment Battery for children.[22]

3.4. Impact of risk factors on speech and language outcome

Impact of developmental risk factors was assessed for the total sample by means of rank correlations (Table 5).

Age at testing was significantly correlated with all four measures of speech and language development. Socioeconomic status was significantly correlated with 2 of the 4 measures. With respect to perioperative risk factors, no significant correlation was found for duration of DHCA. But duration of CPB had a significant influence on TFS and TAAS. Regarding the nonverbal developmental parameters, a significant influence was found on expressive speech measures (TFS or Mayo test), but not on receptive ones (Auditory Closure test and TAAS).

Table 5 *- Developmental and risk factors related to outcome parameters of speech and language*

	TFS		Mayotest score		Auditory closure test score		TAAS	
	SP	*P*[a]	SP	*P*[a]	SP	*P*[a]	SP	*P*[a]
Age at testing (years)	0.45	0.006	0.50	0.002	0.37	0.030	0.87	0.00
Socioeconomic status	-0.07	0.70	-0.38	0.025	-0.36	0.032	-0.04	0.82
Duration of DHCA (min)	-0.09	0.60	-0.10	0.59	-0.05	0.78	-0.04	0.81
Duration of CPB (min)	-0.36	0.032	-0.28	0.11	-0.12	0.51	-0.48	0.004
Motor quotient	0.34	0.046	0.11	0.54	0.13	0.47	0.03	0.87
Intelligence quotient	0.22	0.20	0.55	0.001	0.33	0.053	0.28	0.11
Acquired abilities	0.12	0.51	0.52	0.003	0.31	0.091	0.30	0.10

SP, Spearman correlation coefficient.
DHCA, deep hypothermic cardiocirculatory arrest.
CPB, cardiopulmonary bypass.
[a] *P* value calculated by Spearman correlation coefficient.

4. Discussion

Our data are based on two homogeneous groups of infants with preoperative hypoxemia caused by TOF or with pre-operative cardiac failure caused by a VSD, respectively, in whom preoperative, perioperative, and postoperative care was conducted according to standardized protocols. Follow-up examination of language and speech at mean age 7.6 years after corrective surgery in infancy was performed in 35 children who account for 70% of the operated patients. The outcome data were compared between the subgroup with former hypoxemia (*n* = 19) and the subgroup with former cardiac insufficiency (*n* = 16) in order to test whether the condition of preoperative hypoxemia led to less favorable results in language and speech evaluation. Furthermore, the outcome data were correlated to prospectively assessed factors of the perioperative management, to sociodemographic variables and to recently published results of the actual neurodevelopmental status assessed in the same study group.[14]

4.1. Parental reports and test reults

The present results of parental reports on speech and language history suggest an overall high percentage of reported deficits in language and speech, especially a delayed onset of language development and disorders in the domain of articulation in the whole patient group. Though the prevalence of problems in the Fallot group is elevated on several parameters, significant differences between the subgroups were not found. Our parental report data support the general observation that children after cardiac surgery are found at elevated risk for developmental delay and disorders with respect to language and speech.[2,3,9,10,14,28,29]

The parental reports were corroborated by the follow-up data obtained from standardized speech and language examination. On all measures of oral and speech functions as contained in the Oral and Speech Motor Control Protocol[23] and the Mayo Test of Speech and Oral Apraxia,[24] both groups scored below the average performance or below normal cutoff values of children of the same age as estimated from the validation studies of the respective test instrument. On the other hand, assessment of receptive speech processing (AC[25] and TAAS[26]) demonstrated that deficits of auditory word recognition and phonological awareness are not predominantly prevalent in children after cardiac surgery.

4.2. Differences between subgroups

Regarding the two subgroups who exhibited nearly identical conditions in the preoperative, perioperative and postoperative management and with respect to socioeconomic status, patients with TOF and preoperative hypoxemia have poorer outcomes and are at higher risk for deficits in expressive speech at school age compared to those with VSD and preoperative cardiac insufficiency. Whereas the differences between the subgroups are found borderline with respect to oral structures, evaluation of oral and speech functions demonstrated that Fallot patients have significantly elevated deficits with respect to coordination and control of their organs of articulation.

When the children were individually compared to typical speech development of the same age, more than 50% of the TOF subgroup did not reach normal expressive speech performance in contrast to only 25% of the VSD subgroup. However, with respect to receptive speech and language performance, such a subnormal development was an exception even in the TOF subgroup. Only two children (6%) showed a severe deficit.

Considering the whole cohort, the total functional score (TFS) comprising oral nonspeech functions as well as expressive speech functions was an appropriate measure to separate children with normal and subnormal performance in general, and to identify children with reduced outcome in other domains of speech and language as well as children with poor gross motor function. A similar relationship between motor speech and gross motor dysfunction was previously demonstrated in a cohort of children after neonatal arterial switch operation.[2] As shown for other domains of neurocognitive development,[9,11,13,30-34] dysfunction of expressive speech and language must be considered as an important additional component of developmental impairment in children after palliative or corrective surgery for cyanotic heart disease beyond infancy, especially for those with TOF.

4.3. Impact of developmental parameters and early risk factors

In the whole cohort as well as in both subgroups, higher age at testing went along with better test results. As age-dependent normal values are not available for most of the applied tests, these outcomes could be primarily ascribed to demographic factors. In addition, older children after cardiac surgery during infancy may be able to develop a better compensation of their deficits in language and speech development with time going on.[28] This was supported by our finding that the 29% of all children with consistently poor expressive speech were younger with a mean age of 6.6 years than the 43% of all children with normal speech performance and a mean age of 8.3 years.

In consistency with recent studies providing similar observations in other domains of development,[2,10,14,35] higher socioeconomic status was correlated with better outcome in language and speech.

Perioperative management had an additional and specific influence on the later development. Longer duration of CPB was found inversely related to language and speech results. Furthermore, it was not unexpected that another perioperative risk factor, duration of DHCA could not be identified to significantly influence speech outcome as the conditions of perioperative management were very uniform and the total number of patients was limited.

The children of the present study showed poorer outcome of language and speech in comparison to children after neonatal arterial switch operation for transposition of the great arteries, as assessed with the same test battery at mean age 10.5 years compared to the patients of the present study[2] This suggests that besides protracted cyanosis the global perioperative management with CPB and DHCA during later infancy might be in part responsible for poorer outcome.[14]

Finally, we again demonstrated that dysfunctions in speech were associated with motor, cognitive and academic disabilities[14] as well as with executive attentional dysfunctions[15] in the same patient group. This emphasizes that speech and language disabilities, especially in the field of expressive rather than in the domain of receptive speech items, can be considered as an important indicator of global developmental impairment.[17-19] During the last decade, magnetic resonance anatomical and functional imaging studies of the developing brain have been able to elaborate the time course of maturation processes of different brain areas and their corresponding cognitive functions such as language.[36,37] Whereas the auditory system, associated with the auditory cortex, and the language system in the narrower sense of the word, associated with the sensorimotor cortex (Wernicke and Broca areas), develop in the prenatal or early postnatal period, the speech production system, asscociated with the Broca area, the premotor and primary motor cortex, matures markedly later in infancy and early childhood with respect to synaptogenesis and synaptic pruning. The difference between expressive and receptive speech performance in our children possibly indicates higher sensitivity of still immature prefrontal in contrast to temporal functions of the developing brain after preoperative hypoxemia as well as due to risk factors of the perioperative management.[38,39]

In conclusion, chronic preoperative hypoxemia rather than chronic preoperative heart failure is associated with unfavorable outcome of language and speech with special respect to expressive functions. Age at testing, socioeconomic status, and duration of CPB influence the test results. Long-term outcome in language and speech in general can be considered as a sensitive indicator of child development after cardiac surgery.

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