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ORIGINAL ARTICLE

Global intellectual ability and adaptive functioning in children with FASD with and without sentinel facial features

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ABSTRACT

Background: Fetal Alcohol Spectrum Disorder (FASD) is a neurodevelopmental disorder characterized by cognitive and adaptive impairment. FASF can be presented or not with sentinel facial features (SFF). The presence of such SFF have been positively correlated with cognitive impairment in children with FASD.

Objectives: The current study explores difference in global intellectual functioning and how cognition affects adaptive behavior in children with and without SFF.

Methods: A total of 88 children and adolescents (55 males, 33 females) with confirmed FASD diagnosis were included in the study, of which 16 had sentinel facial features. Childrens' neuropsychological functioning was assessed using the Wechsler Intelligence Scale for Children (WISC-V) and The Behavioral Assessment of the Dysexecutive Syndromes for Children (BADS-C). Adaptive behavior was explored through the Adaptive Behavior Assessment System (ABAS-3).

Results: Children with SFF performed more poorly in tasks assessing processing speed ($t = 2.495$, $t = .020$) and executive functioning ($t = 4.147$, $t = .001$). Those children also had lower IQ scores than children without SFF ($t = 2.658$, $t = .016$). BADS-C overall scaled score was related to three of the four domains of the ABAS scale (conceptual, social, and practical) but only in the group of FASD children without SFF (B = 0.547, $t = .020$; B = 0.544, $t = .049$; B = 0.431, $t = .040$, respectively).

Conclusions: The present study founds poorer cognitive outcomes in children who have FASD with sentinel facial features. In children without SFF, stronger executive functioning is also related to significantly stronger reported conceptual, social, and practical adaptive behaviors. Better understanding of cognitive and adaptive functioning in children with FASD may help in the design of tailored evidenced-based interventions.

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Introduction

Alcohol consumed during pregnancy passes directly through the placenta into the fetus' bloodstream and amniotic fluid (1), causing significant life-long teratogenic effects, including functional and developmental abnormalities (2,3).

Fetal Alcohol Spectrum Disorder (FASD) is an umbrella term used to describe the "variety of syndrome manifestations characterized by physical and mental abnormalities, as well as behavioral and learning difficulties" caused by prenatal alcohol exposure (4). FASD is distinctive to other neurodevelopmental disorders, in the sense that it is entirely preventable (5,6). In the United States and Western Europe alone, although the exact figures are unknown and may vary across the world, a 1–5% FASD prevalence has been estimated (7). A recent prevalence study of primary school children in Greater Manchester UK, found that 1.8% of the children participating had FASD. This increased to 3.6% when including children with "possible" FASD (8). The study was ground-breaking in terms of demonstrating the significant numbers of children affected by this condition.

FASD is a neurodevelopmental disorder (9,10) that can present or not with sentinel facial features (11), which are characteristic anatomical facial features in some of the patients with FASD. According to SIGN Guidelines, only 10% of the children with FASD have

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sentinel facial features, and therefore their presence or absence is not as significant diagnostically as the neurocognitive and neurodevelopmental aspects of the diagnosis. Children with FASD may display a specific pattern of three defining sentinel facial features (SFF) as evidence of prenatal alcohol exposure (PAE). Only when all three features present together are this specific to PAE. The three SFFs include short palpebral fissure (<3th percentile, according to SIGN Guidelines) and lip and philtrum length and shape at grade 4 or 5 on the University of Washington Lip and Philtrum Guide (Lip-Philtrum Guides (9). Epicanthal folds, flat nasal bridge, anteverted nares, ear shape and palmar creases are also sometimes observed; however, these are not accepted diagnostic signs (12). When all three SFFs are present, FASD diagnosis may be made without documented information of PAE (9). While the majority of children with FASD do not have SFF (90%), presence of SFF has been described as relating to poorer cognitive functioning in several studies (13,14).

PAE can cause significant cerebrum, cerebellum, and congenital abnormalities, as well as atypical central nervous system development, often resulting in lower global intellect, poor attention skills, and slower reaction times (3). Visual and auditory attention are both often impaired, resulting in deficits in verbal learning particularly (7,15). Hot and cold executive functioning (attention, adaptive functioning, planning, and flexibility of thinking and response inhibition) and general cognitive ability (behavioral, social, and emotional regulation) are also affected, all of which are necessary for goal directed behavior and social success. Deficits in speech and language development, including receptive, expressive, and syntactical ability can further negatively affect upon a child's overall academic functioning. Children with FASD often present with deficits in social cognition, including problems with social interaction, social communication, and general social competence (7). The far-reaching range of neurocognitive and neurodevelopmental impairment observed in children with FASD has significant implications for their ability to manage daily life, and therefore considerably impacts upon adaptive functioning life (16,17). The adaptation process in daily life supposes a continuous process of construction based on learning from experiences and assessment of results, for which adequate neurocognitive functioning is essential.

Research suggests that more dysmorphic features in children with FASD are related to poorer IQ and lower overall cognitive capacity as these may relate directly to damage to certain parts of the developing brain (13,18,19). Specifically, the presence of the three specific anthropometric facial measures in children with FASD have been positively correlated with cognitive impairment (14,20).

The current study explores difference in global intellectual functioning and how cognition affects adaptive behavior in children with and without SFF.

Method

Participants

In the current cross-sectional study, we examined data from children attending the Center for FASD for assessment. The center, located in Suffolk (UK), is a private assessment clinic. All children referred were presenting with complex behavioral and neurodevelopmental difficulties, with a history of confirmed PAE. FASD assessment and diagnosis was made according to SIGN diagnostic Guidelines, including establishing whether SFF were present. Lip and Philtrum grade were assigned according to the University of Washington Lip/Philtrum guide, which has been a method extensively assessed and validated (21) and palpebral fissures were measured directly and verified using the Facial Photographic Analysis Software from the University of Washington.

The children underwent three key assessment sessions face-to-face, including pediatric assessment, neuropsychological assessment with the clinical psychologist, and speech and language assessment with the speech and language therapist, all of whom have expertise in FASD assessment and diagnosis. The pediatrician took a developmental history from the parents, and the clinical psychologist took a more detailed history of the child's behavior in different contexts, The child's school was also consulted and parents and teachers were asked to complete standardized questionnaires. If FASD diagnosis was confirmed, the child's data was entered into the current study and classified as FASD with SFF, or FASD without SFF.

As part of the neuropsychological assessment, the Wechsler Intelligence Scale for Children (WISC-V) (22) and The Behavioral Assessment of the Dysexecutive Syndromes for Children (BADS-C) (23) were used.

The WISC-V is a measure of global intellectual functioning for children from 6 to 16 years of age with five key composite scores retrieved from subtest results; 1) The Verbal Comprehension Index, which indicates how well the patient can respond to questions about the social world, understand, and process verbal information, think and reason with words, and express himself; 2) The Visual Spatial Index, which indicates a child´s capacity to understand, reason, problem-solve, and remember spatial relations between objects; 3) The Fluid Reasoning Index, which provides an understanding of the capacity to reason and solve novel problems, independent of any pre-acquired knowledge; 4) The Working Memory Index, which indicates the ability to retain and manipulate information within the short-term memory store; 5) The Processing Speed Index measures speed of mental problem solving, attention, and eye-hand co-ordination. Finally, a Full-Scale IQ is derived from a combination of the above composite scores and is a means of understanding an individual's general level of cognitive and learning ability. The full-scale IQ score is only valid where there is no significant variance across composite scores.

The BADS-C is a good measure of "cold" executive functioning processes such as planning, sequencing, problem solving, switching, inhibitory control, and initiation processes. It consists of six discrete subtests and provides an overall quantitative score of executive functioning and a qualitative interpretation of this score.

Adaptive functioning was assessed using the Adaptive Behavior Assessment System (ABAS-3) (24) which is made up of four composite scores: 1) the General Adaptive Composite, that summarizes performance across all skill areas, 2) Conceptual, that describes communication abilities including academic performance and functional skills, 3) Social, indicating the ability to plan and engage in leisure activities, to interact and maintain appropriate friendships and to understand emotions, and 4) Practical, that provides a specific picture of the child with regard to his independence skills, his ability to keep himself safe and to take care of his personal hygiene. The parent/carer scale version was used to assess adaptive functioning.

Procedure

Ethical issues

All participants gave written informed consent to participate in the study and in the case of participants under 18, their parents/legal guardian also signed the informed consent form. The study protocol was approved by the corresponding Ethics Committee and all the research conducted in this study meets the "Ethical principles of psychologists and code of conduct" (25).

Statistical analysis

For descriptive purposes, continuous variables were presented as mean and standard deviation (SD); percentages were used to describe categorical variables. Sociodemographic characteristics and cognitive performance of the two groups were compared through the independent-samples t-test (Student's t-test) for continuous variables and contingency tables for categorical variables ($\chi 2$ or Fisher's exact test if $n \leq 5$ per cell). For continuous variables, normal distribution was assumed after the application of Shapiro-Wilks test and the qq-plots. A multivariate analysis (linear regression model) was performed in order to explore the influence of cognitive impairment in adaptive functioning; Such analysis included cognitive domains as independent variables and the score of the subtests of the ABAS test as dependent ones. For all the analyses, P-values below $n < .05$ were considered statistically significant. Age was considered as a covariable of the model.

Data were analyzed using the statistical packages IBM SPSS Statistics 23 and R version 4.1.2.

Results

A total of 88 children and adolescents with confirmed FASD diagnosis were included in the study (55 males and 33 females), of which 16 had sentinel facial features (18.2%).

Table 1 summarizes the main characteristics of the study participants. Participants' ages ranged from 3 to 19 years and they were all adopted children. No significant differences in sociodemographic characteristic were found between children with or without sentinel facial features

In relation to cognitive functioning, children with SFF performed more poorly in tasks assessing processing speed and executive functioning. Those children also had lower IQ scores than children without SFF. The scores of the cognitive indexes of the WISC-V and the BADS-C obtained in both groups are shown in Table 2.

In order to explore how differences in cognitive functioning may influence adaptive functioning children with FASD, we built linear regression models with these three cognitive domains, which showed differences

Table 1. Characteristics of the sample ($N = 88$).

	FASD without SFF	FASD with SFF	Statistic	p value
n (%)	72 (81.80)	16 (18.20)		
Sex, male, n (%)	44 (61,10)	11 (66,70)	$X^2 = 0.08$	.775
Age, in years, mean (SD)	9.97 (3.61)	8.56 (3, 05)	t = 1.61	0,119
Birth Weight in kg, mean (SD)	2.38 (0,64)	2.08 (0,70)	t = 1.12	.285

FASD: Fetal Alcohol Spectrum Disorder.
SFF: Sentinel Facial Features.

Table 2. Comparison between FASD children with or without sentinel facial features (SFF) in cognitive indexes of the WISC-IV and BADS-C.

	FASD without SFF (*N* = 72)		FASD with SFF (*N* = 16)			
	Mean	SD	Mean	SD	Statistic	*p* value
Verbal Comprehension	86.97	16,32	78.71	16,10	t = 1.72	0,101
Visual Spatial	86.28	13.02	82.50	1169	t = 1.06	.301
Fluid Reasoning	84.55	14.21	81.71	9.35	t = 0.91	0,370
Working Memory	81.26	13.95	76.71	9.93	t = 1.41	.170
Processing Speed	84.62	19.00	73.50	12.81	t = 2.50	.020
Full scale IQ	83.61	13.62	74.00	9.42	t = 2.66	.016
BADS-C Overall scaled score	67.11	18.31	51.25	4.72	t = 4.15	.001

FASD: Fetal Alcohol Spectrum Disorder.
SFF: Sentinel Facial Features.
IQ: Intelligence Quotient.
BADS-C: The Behavioural Assessment of the Dysexecutive Syndromes for Children Scales.

between groups as independent variables and the domains of the ABAS test as dependent ones. Results are shown in Table 3. As this table demonstrates, BADS-C overall scaled score was related to three of the four domains of the ABAS scale (conceptual, social and practical) but only in the group of FASD children without SFF.

Discussion

The present study supports the current literature base, which suggests poorer cognitive outcomes in children and young people who have FASD with sentinel facial features. The findings obtained suggest that children and young people who have FASD with SFF have poorer cognitive performance in terms of their processing speed, and in their “cold” executive functioning skills than those who have FASD without SFF. Full-scale IQ scores were also found to be lower in those with SFF. In children and young people without SFF, stronger executive functioning (albeit generally still impaired) is also related to significantly stronger reported conceptual, social, and practical adaptive behaviors.

A strength of our study is the large sample of children with FASD included with an important representation of children with SFF (18.2%). According to the Kautz-Turnbull meta-analysis (26) there could exist an smaller effect size in active case detection than in clinical cohorts, so our results may even be underestimated.

Slower processing speed has been described in children with FASD previously (27). However, to date, this is the first article to differentiate between children and young people with FASD who have, and do not have, sentinel facial features. A significant difference was found, with children and young people who have sentinel facial features, demonstrating slower processing speed overall.

The relatively stronger executive functioning skills of children and young people with FASD without SFF is also associated with better adaptive behavior and functioning. This provides further evidence of the association between executive function and adaptive behavior, and is consistent with the findings of Kautz-Turnbull et al., who in their meta-analytical review, also found that impaired executive functioning in children and young people with FASD modulates the development of certain important adaptive capacities for daily life. They also found no relationship between executive and adaptive function, and IQ (26). This supports the fact that most children and young people with FASD (with

Table 3. Linear regression models to explore the influence of cognitive domains in adaptive functioning.

	FASD with SFF								FASD without SFF							
	ABAS General		ABAS Conceptual		ABAS Social		ABAS Practical		ABAS General		ABAS Conceptual		ABAS Social		ABAS Practical	
	B	*p*	B	*p*	B	*p*	B	*p*	B	*p*	B	*p*	B	*p*	B	*p*
Processing speed	0.49	.189	0.07	.898	−0.11	.856	0.36	.450	0.09	.293	−0.19	.393	−0.23	.379	−0.12	.519
IQ	0.92	.103	−0.20	.852	−0.47	.692	−0.10	.916	0.11	.391	0.48	.150	0.31	.417	0.23	.382
BADS-C overall	1.49	.182	1.90	.681	3.81	.464	3.60	.299	0.04	.600	0.55	.020	0.54	.049	0.43	.040

B: regression coefficient.
p: *p*-value.
FASD: Fetal Alcohol Spectrum Disorder.
SFF: Sentinel Facial Features.
IQ: Intelligence Quotient.
BADS-C: The Behavioural Assessment of the Dysexecutive Syndromes for Children Scale.
ABAS: Adaptive Behaviour Assessment System.

or without SFF) have near normal IQ, but have executive functioning skills (and therefore adaptive skills) that are significantly below this level. This is a key component to the "spikiness" in the neurodevelopmental profiles of children with this condition.

Better understanding of cognitive and adaptive functioning in children with FASD may help in the design of tailored evidenced-based interventions to support scaffolding around poor adaptive behavior for those affected by FASD, in order to ensure as successful and safe a transition to adulthood as possible (17). It has, for example, been previously shown that altering and tailoring parenting approach for children who have FASD, can facilitate their ability to function well in different environments, including optimally supporting executive functioning skills (28).

In conclusion, the present study find poorer cognitive and adaptive outcomes in children who have FASD with sentinel facial features. Conversely, in children who have FASD without SFF, stronger executive functioning is related to significantly stronger reported conceptual, social, and practical adaptive behaviors.

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