



Social Behavior in RASopathies and Idiopathic Autism

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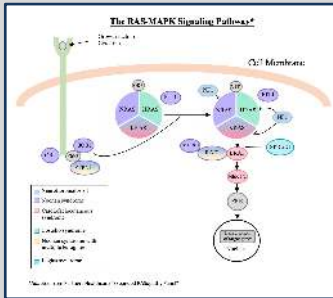
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Introduction

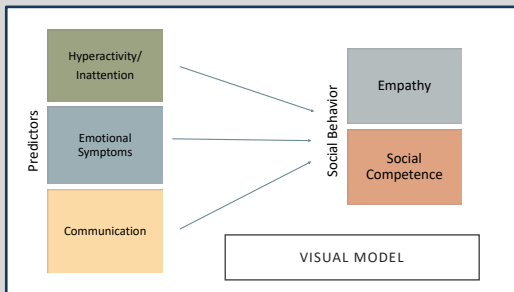
- Gene mutations within the RAS-MAPK signaling cascade cause cardiofaciocutaneous (CFC) syndrome, Costello syndrome, Noonan syndrome (NS), neurofibromatosis type 1 (NF1) and related disorders.
- Children with these "RASopathy" syndromes and children with non-syndromic autism spectrum disorder (idiopathic ASD) are both at increased risk of social problems, but few studies have compared social behavior in these conditions (Adviento et al., 2014).
- Some literature indicates that children with RASopathies may exhibit relatively strong empathetic or prosocial behaviors (Noll et al., 2007).



Objectives

Objective 1: To evaluate and compare the presence of empathy and social competence in children and adolescents with RASopathies and idiopathic ASD

Objective 2: To determine which psychological factors predict scores on measures of empathy and social competence for children in each group



Participants & Methods

- Cross-sectional, survey-based investigation
- Parents and caregivers of 180 children with RASopathies (NF1, NS, CFC syndrome, Costello syndrome) and 97 children with idiopathic ASD
- Rated the presence of empathetic behavior and social competence as well as symptoms of hyperactivity/inattention, emotional problems, and communication skills. Medical and family history was also ascertained.

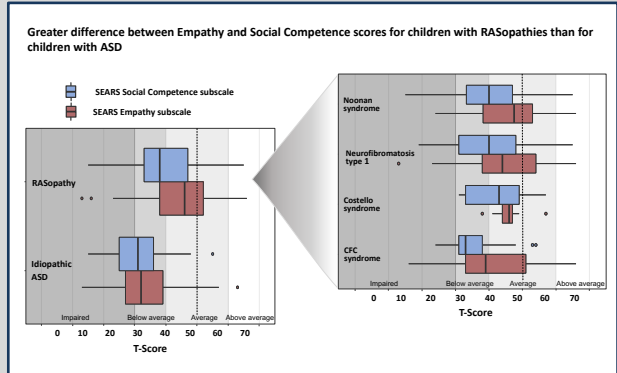
Measures	
Background Questionnaire	Demographic Information
GO4KIDD5	Communication • Expressive • Receptive
Strengths & Difficulties Questionnaire (SDQ)	• Emotional Symptoms • Hyperactivity/Inattention
Social Emotional Assets & Resilience Scales (SEARS)	• Empathy • Social Competence

Participant demographics	RASopathies					Idiopathic ASD (n=110)
	NF1 (n=71)	NS (n=73)	CFCs (n=41)	CS (n=18)	All RASopathies (n=203)	
Age of Child (mean)	9.84	9.2	8.79	10.95	9.5	10.41
Gender (% Male)	51*	51	42	50	49	73*
Race (%)						
American Indian or Alaska Native	0	1	2	0	1	6
Asian	3	6	7	11	5	5
Black or African American	6	4	5	0	4	5
Native Hawaiian or Other Pacific Islander	1	1	0	11	2	2
White	92	95	90	94	93	88
Other	1	1	5	11	3	6
Ethnicity (%)						
Hispanic/Latinx	18	6	7	6	10	15

*One participant did not identify with male or female gender

Results

RASopathy	Idiopathic ASD
Relatively intact empathy despite higher risk for social competence problems. This was true even for children with autism diagnoses.	Similar impairment across social competence and empathy
Stronger communication skills are associated with increased empathetic behavior	
Problems with social competence were more associated with emotional challenges and hyperactive-impulsive behaviors	Problems with social competence were more associated with communication difficulties



Scores on social behavior measures for children with comorbid RASopathy+ASD as compared to idiopathic ASD

Measure	IASD				RAS+ASD			
	n	Mean	(SD)	% Clinical Impairment (≥2 SD below mean)	n	Mean	(SD)	% Clinical Impairment (≥2 SD below mean)
Social Emotional Assets & Resilience Scales (SEARS)								
Social Competence	88	30.49	(7.64)	48	20	30.65	(8.29)	45
Empathy	87	32.99	(9.39)	40	20	35.60	(8.56)	25
Strengths & Difficulties Questionnaire (SDQ)								
Emotional Symptoms	97	1.32	(1.29)	29	22	1.88	(1.65)	41
Conduct Problems	97	0.71	(1.10)	17	22	0.66	(0.91)	9
Hyperactivity/Inattention	97	1.59	(1.03)	46	22	1.86	(1.00)	50
Peer Relationship Problems	97	2.21	(1.11)	58	22	1.80	(1.34)	55
Prosocial Behavior	97	1.79	(1.24)	44	22	1.53	(0.96)	36

Conclusions

- Empathetic behavior is frequently observed in children with RASopathies, thus differentiating the social problems experienced by these children from those experienced by children with iASD
- The patterns of psychological factors associated with different areas of social strength and weakness differ among children with RASopathy and iASD
- Thus, interventions for social difficulties should target the specific social strengths and weaknesses experienced as well as the developmental and neuropsychological factors underlying the social difficulties

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