

## Factors associated with self-reported functional health status in a multi-institutional cohort of young adults with Interrupted Aortic Arch

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**Objectives:** Improved survival after congenital heart surgery has led to increasing interest in functional health status (FHS) of patients as they transition to adulthood. We sought to identify factors associated with self-reported FHS of young adults with repaired IAA.

**Methods:** Follow-up in 2010 of survivors (aged 13-24 years) from a 1987-1997 inception cohort of neonates with IAA included completion of FHS questionnaires [Child Health Questionnaire-CF87 (age <18, n=51) or the Short Form-36 (age ≥18, n=66)], and a questionnaire about features of 22q11 deletion syndrome (DS) (n=141 survivors), as few subjects had had genetic testing. Patient characteristics (including 22q11DS features), medical history, and psychosocial factors associated with FHS domains were sought using multivariable linear regression analyses.

**Results:** Domain scores were significantly higher than norms in 2/9 CHQ-CF87 and 4/10 SF-36 domains, and only lower in the physical functioning domain of the SF-36 (table). Factors most commonly associated with lower scores were those suggestive of features of 22q11DS (low calcium levels, recurrent childhood infections, genetic testing/diagnosis, abnormal facial features, hearing deficits), the presence of behavioral and mental health problems, and a higher number of procedures. Poor FHS scores were less commonly associated with specific anatomy, higher number of medications, lower family income, lower weight and age at the index repair, shorter procedure free interval, and having other medical problems. Depending on the FHS domain, factors explained from 10% up to 70% of score variability ( $R^2=0.10-0.70$ , adj- $R^2=0.09-0.66$ ). Of note, FHS was minimally related to IAA morphology and repair type.

**Conclusions:** Morbidities related to 22q11DS, psychosocial and recurrent medical problems affect

Table. Comparison of IAA Patients to Normative Data

CHQ-CF87 Domain	IAA patients <18y Mean (SD)	Normal 10-15 year olds Mean (SD)	P
Global Health (GGH)	77.2 (17.2)	-	-
Physical Functioning (PF)	86.6 (15.6)	88.8 (14.0)	.3
Role/Social Limitations – Emotional (RE)	85.6 (20.7)	85.9 (21.0)	.9
Role/Social Limitations – Behavioral (RB)	88.0 (25.3)	86.5 (21.5)	.7
Role/Social Limitations – Physical (RP)	92.3 (16.7)	88.3 (21.0)	.1
Freedom from Bodily Pain (BP)	87.8 (19.3)	74.4 (23.1)	<.0001
Behavior (BE)	73.0 (16.5)	76.6 (14.6)	.1
Global Behavior (GBE)	71.0 (27.6)	-	-
Mental Health (MH)	77.6 (15.2)	72.7 (16.0)	.03
Self Esteem (SE)	79.3 (15.6)	81.8 (15.8)	.3
General Health Perceptions (GH)	64.5 (14.8)	66.4 (14.6)	.4
Family Activities (FA)	80.0 (23.8)	-	-
Family Cohesion (FC)	73.9 (22.6)	-	-
SF-36 Domain	IAA patients ≥18y Mean (SD)	Normal 18-24 year olds Mean (SD)	P
Physical Component Summary (PF/RP/BP/GH)	52.4 (7.5)	53.5 (9.2)	.3
Mental Component Summary (VT/SF/RE/MH)	49.3 (12.0)	46.1 (13.3)	.04
Physical Functioning (PF)	50.7 (8.0)	53.2 (9.7)	.02
Role-Physical (RP)	50.9 (8.6)	52.8 (9.6)	.09
Freedom from Bodily Pain (BP)	55.9 (8.0)	52.0 (10.6)	.0002
General Health (GH)	49.2 (10.9)	49.7 (11.8)	.7
Vitality (VT)	52.6 (11.3)	47.0 (11.7)	.0002
Social Functioning (SF)	49.1 (10.0)	49.2 (12.3)	.9
Role-Emotional (RE)	49.0 (11.8)	49.8 (12.5)	.6
Mental Health (MH)	50.7 (11.8)	46.9 (13.0)	.01

\* Note that a higher score for any given domain denotes better functional health status as related to that category.

FHS in IAA survivors, and dominate over cardiac history. Nonetheless, these survivors generally perceive themselves to have higher FHS than their peers – a seemingly paradoxical association that may reflect known phenomena (response shift, disability paradox, sense of coherence). Evaluation and surveillance/strategies aimed at definitive surgical treatment, mental health, and genetic issues might be an important program component of cardiac care in the transition from adolescence through early adulthood. Ongoing assessment of FHS in this cohort will be required to detect deteriorations related to increasing complexity and stress associated with mature adult roles.