The Association Between Age at Diagnosis and Health-Related Quality of Life for Patients with ANCA-Associated Vasculitis

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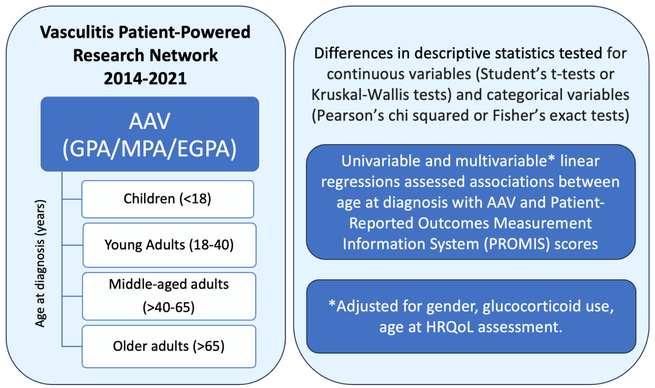
BACKGROUND

- ANCA-associated vasculitis (AAV) can occur throughout the lifespan and impacts health-related quality of life (HRQoL)
- It is unknown whether this impact varies based on age at diagnosis, from childhood through adulthood
- This study examined the relationship between age at diagnosis of AAV and HRQoL

METHODS



GPA, granulomatosis with polyangiitis; *MPA*, microscopic polyangiitis; *EGPA*, eosinophilic GPA.

RESULTS

- 2009 patients: 1351 (67.2%) GPA, 301 (15.0%) MPA, 357 (17.8%) EGPA. (Table 1)
- Female predominance decreased with age at diagnosis.
- Those diagnosed at younger ages had longer time since diagnosis at last PROMIS assessment
- All cohorts had impaired HRQoL in every domain, especially anxiety (Figure 1)
- Use of glucocorticoids impacted HRQoL
- When adjusted, the PROMIS scores for those diagnosed at younger ages were:
  - *better* for fatigue, sleep disturbance, depression, pain interference, and satisfaction with participation in social roles
  - the *same* for physical functioning
  - *worse* for anxiety, compared to those diagnosed at older ages (Table 2)

Table 1. Characteristics of patients with ANCA-associated vasculitis based on age at diagnosis.

	Age at Diagnosis					
	Child *<18 years old*	**Young Adult** *18-40 years old*	**Middle-Aged** *>40-65 years old*	**Older Adult** *>65 years old*	**All**	
	N = 110	N = 507	N = 1172	N = 220	N = 2009	
Characteristic	*Mean (standard deviation) or n (percentage)*					***p*-value***
Age at diagnosis (years)	13.3 (3.7)	30.7 (6.6)	53.5 (6.8)	69.9 (3.7)	47.4 (15.9)	< 0.001
Sex (male)	21 (19.1%)	125 (24.7%)	368 (31.4%)	83 (37.7%)	597 (29.7%)	0.008
Disease category						< 0.001
GPA	78 (70.9%)	375 (74.0%)	761 (64.9%)	137 (62.3%)	1351 (67.2%)	
MPA	21 (19.1%)	49 (9.7%)	177 (15.1%)	54 (24.5%)	301 (15.0%)	
EGPA	11 (10.0%)	83 (16.4%)	234 (20.0%)	29 (13.2%)	357 (17.8%)	
Time since diagnosis at quality of life assessment (years)	12.6 (13.3)	11.3 (10.5)	8.1 (6.5)	5.1 (4.7)	8.8 (8.3)	< 0.001

GPA, granulomatosis with polyangiitis; *MPA*, microscopic polyangiitis; *EGPA*, eosinophilic granulomatosis with polyangiitis. Unknown or missing responses for each score were not included. *Statistical significance was set at 0.05.

Table 2. Differences in PROMIS scores among patients with ANCA-associated vasculitis based on age at diagnosis.

		Differences‡ by Age at Diagnosis		
PROMIS Measure	**N**	**Child vs Young Adult**	**Child vs Middle Age**	**Child vs Older Age**
Lower score indicates better health-related quality of life				
Fatigue	1876	-3.72 (-6.22,-1.22)	-6.13 (-9.05,-3.21)	-6.65 (-10.32,-2.98)
Sleep Disturbance	1276	-3.97 (-6.37,-1.57)	-6.57 (-9.39,-3.75)	-5.22 (-8.85,-1.60)
Depression	1878	-3.69 (-5.88,-1.51)	-6.44 (-8.99,-3.90)	-7.18 (-10.37,-3.98)
Anxiety	1875	3.28 (1.37,5.19)	5.81 (3.59,8.04)	7.57 (4.78,10.36)
Pain Interference	1877	-2.60 (-4.93,-0.26)	-4.32 (-7.03,-1.6)	-2.97 (-6.39,0.44)
Higher score indicates better health-related quality of life				
Social Satisfaction	1277	3.27 (0.43,6.10)	8.11 (4.76,11.46)	8.01 (3.68,12.35)
Physical Functioning	1884	-0.03 (-2.11,2.04)	1.48 (-0.93,3.90)	1.60 (-1.43,4.62)

PROMIS, Patient-Reported Outcomes Measurement Information System. *Statistical significance was set at 0.05. ‡Differences assessed via multivariable linear regression adjusting for gender, glucocorticoid use, and age at time of quality-of-life assessment. If confidence interval includes zero, the difference is not significant. A positive estimated difference in scores represents a higher score for those diagnosed in childhood compared to the reference group, while a negative difference represents a lower score.

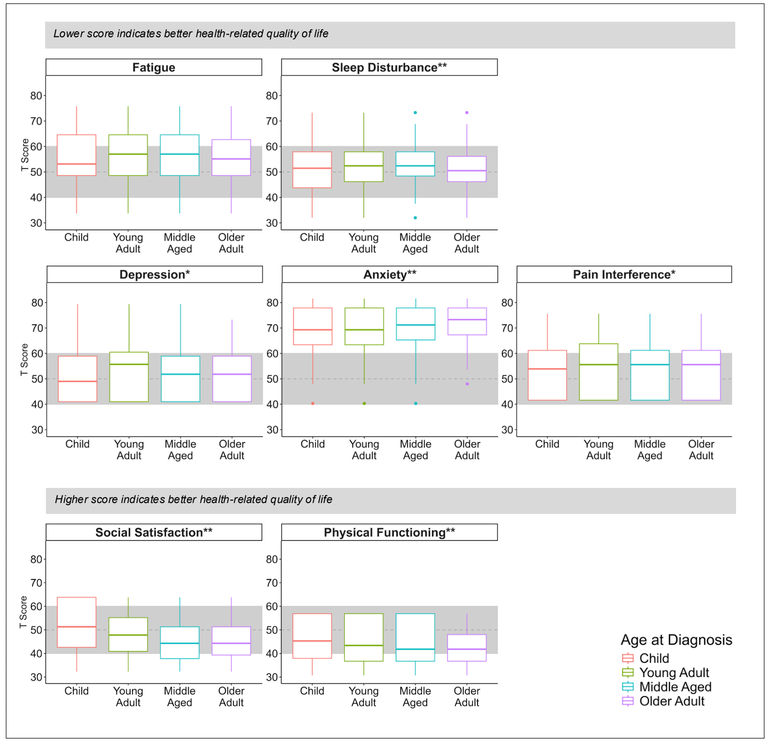


Figure 1. PROMIS T-Scores Based on Age at Diagnosis with ANCA-associated vasculitis. Boxes represent 25th to 75th percentile T scores; middle line=median T score. Dashed gray line=mean score of referent general pediatric population (50), with one standard deviation above and below represented in gray. Better health-related quality of life is represented by a lower score in the top five measures and a higher score in the bottom two measures. Statistical significance indicates at least one group's median T score is different from another.
*statistical significance at $p < 0.05$; **statistical significance at $p < 0.01$.

CONCLUSIONS

- Patients with AAV have impaired HRQoL, especially related to anxiety.
- Age at diagnosis with AAV is associated with differences in HRQoL.
- Compared to patients diagnosed with AAV at older ages, those diagnosed at younger ages have worse HRQoL related to anxiety and better HRQoL related to fatigue, sleep disturbance, depression, pain interference, satisfaction with participation in social roles, and physical functioning.
- Understanding mechanisms by which age at diagnosis impacts HRQoL will inform development of targeted interventions for people with AAV of all ages.

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