EFFECT OF LIVER DISEASE, NEUROLOGICAL DISEASE AND MENTAL HEALTH ISSUES ON QUALITY OF LIFE IN PATIENTS WITH WILSON DISEASE

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Background Wilson Disease (WD) can result in a number of physical and mental health issues due to copper toxicity affecting the liver and brain. It is no longer acceptable to look at survival alone as an outcome measure in chronic disease. Our aim was to a) assess quality of life (QOL) in WD b) assess the relationship between mental health QOL (M-QOL) and physical health QOL (P-QOL) and severity of liver and neurological disease and mental health QOL issues (major depressive disorder (MDD) and cognitive impairment).

Methods Adult WD patients in the WD registry study (n=52), were evaluated at enrollment over 1.5 y using questionnaires and administered exams assessing QOL (SF-12), cognition (MoCA) and mood (MINI-7). Patients also underwent hepatology and neurological assessments (UWDRS).

Results Adult patients with WD had lower M-QOL scores compared to P-QOL scores median 50.2 (range 19.3–62.2) vs. 56.0 (range 23.9–64.7), p=0.0018 indicating that the burden of mental health issues in WD is greater than that of physical health issues on quality of life.

Eleven patients had cirrhosis based on review of imaging, APRI and Fib4 scores. There was no significant difference in M-QOL scores in patients with cirrhosis vs. those without (median 54.5 (range 26.2–59.7) vs. 46.5 (range 19.3–62.2), p = 0.11. Similarly, there was no significant difference in P-QOL in patients with cirrhosis vs. those without (median 55.7 (range 44.4–59.9) vs. 56.3 (range 23.9–64.7), p = 1.00. In those with cirrhosis, higher Child-Pugh scores were associated with a worse P-QOL (r=-0.84, p=-0.0011) and M-QOL (r=-0.60, p=0.0488).

Patients with lifetime MDD (n=22) had worse M-QOL scores compared to those without MDD (median 42.3 vs 54.6, p<0.001). We found no significant difference for those with MDD in P-QOL scores in those with MDD compared to without (median 53.7 vs 54.0, p=0.39). We did not find an association with impaired cognition and QOL scores.

The P-QOL scores have a moderate negative association with the neurological UWDRS II Score (r=-0.44, p=0.001), UWDRS III Score (r=-0.42, p= 0.002) and total UWDRS score (r=-0.44, p=0.001). There are no associations with M-QOL and neurological UWDRS scores.

Conclusions While overall QOL in WD is affected by both mental and physical health, patients with WD have worse M-QOL than P-QOL scores and mental health issues may affect WD patient’s QOL independent of their degree of liver or neurologic disease. Multivariate regression will be performed to evaluate if mental health issues are independently associated with QOL.