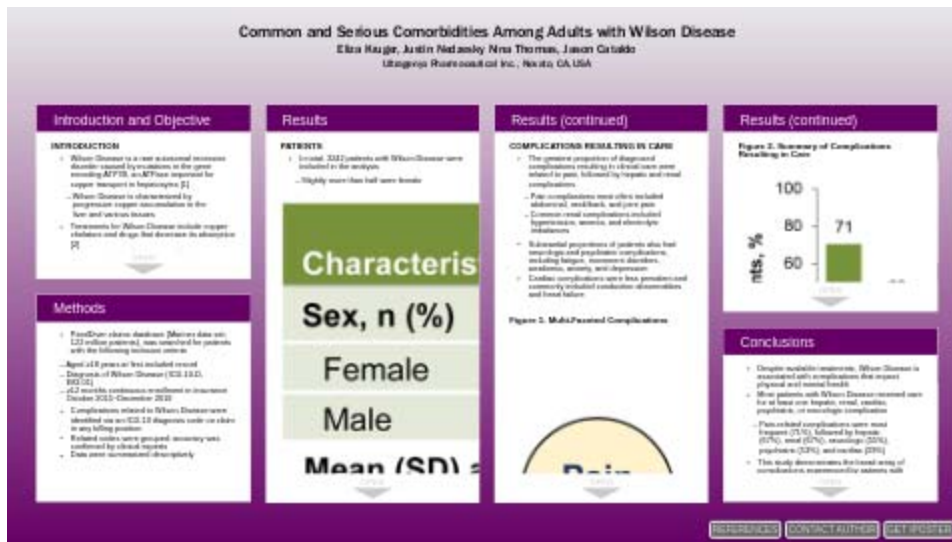


Common and Serious Comorbidities Among Adults with Wilson Disease



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PRESENTED AT:



INTRODUCTION AND OBJECTIVE

INTRODUCTION

- Wilson Disease is a rare autosomal recessive disorder caused by mutations in the gene encoding ATP7B, an ATPase important for copper transport in hepatocytes [1]
 - Wilson Disease is characterized by progressive copper accumulation in the liver and various tissues
- Treatments for Wilson Disease include copper chelators and drugs that decrease its absorption [2]
- If untreated, complications resulting from copper accumulation include liver cirrhosis, psychiatric and neurologic disease (eg, tremor and affective disorder), and dysfunction of other organs (eg, heart and kidneys) [1,3]
- Progressive complications of Wilson Disease have not been quantified in a large cohort

OBJECTIVE

- This retrospective cohort study investigated the most common chronic complications in adults with Wilson Disease

METHODS

- PearlDiver claims database (Mariner data set; 122 million patients), was searched for patients with the following inclusion criteria
 - Aged ≥ 18 years at first included record
 - Diagnosis of Wilson Disease (ICD-10-D-E83.01)
 - ≥ 12 months continuous enrollment in insurance
October 2015–December 2019
- Complications related to Wilson Disease were identified via an ICD-10 diagnosis code on claim in any billing position
- Related codes were grouped; accuracy was confirmed by clinical experts
- Data were summarized descriptively

RESULTS

PATIENTS

- In total, 3242 patients with Wilson Disease were included in the analysis
 - Slightly more than half were female

Characteristic	Patients N=3242
Sex, n (%)	
Female	1743 (54)
Male	1499 (46)
Mean (SD) age, years	46.4 (16.3)
Mean (SD) duration of enrollment, years	3.4 (0.8)
Type of insurance plan, n (%)	
Commercial	2345 (72)
Medicare	447 (14)
Medicaid	320 (10)
US Department of Veterans Affairs	49 (2)
Not available	81 (2)
Region, n (%)	
South	1163 (36)
Northeast	824 (25)
Midwest	755 (23)
West	491 (15)
Unknown	*(<1)

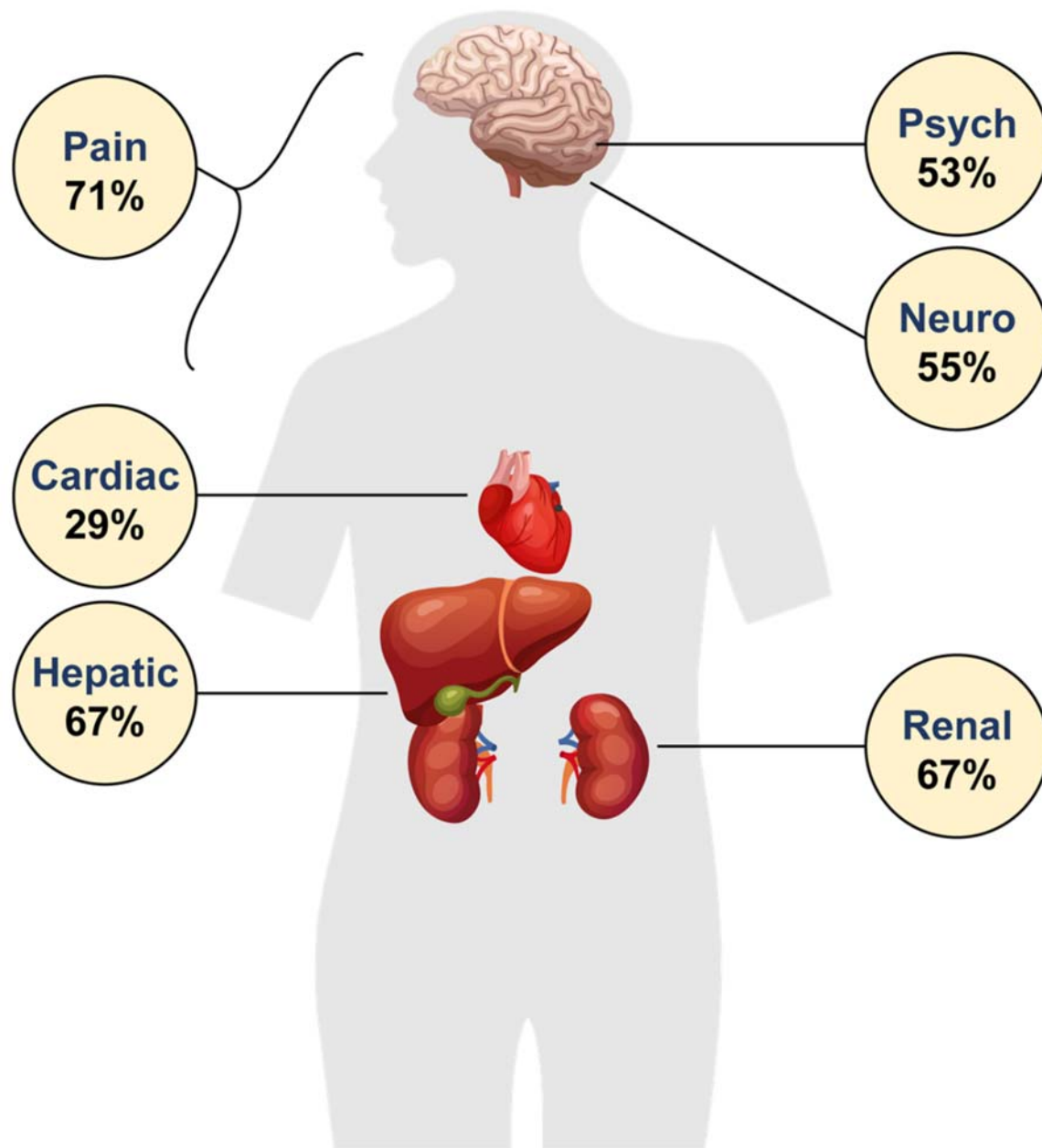
*Patient counts of 1–10 were not reported for privacy reasons.

RESULTS (CONTINUED)

COMPLICATIONS RESULTING IN CARE

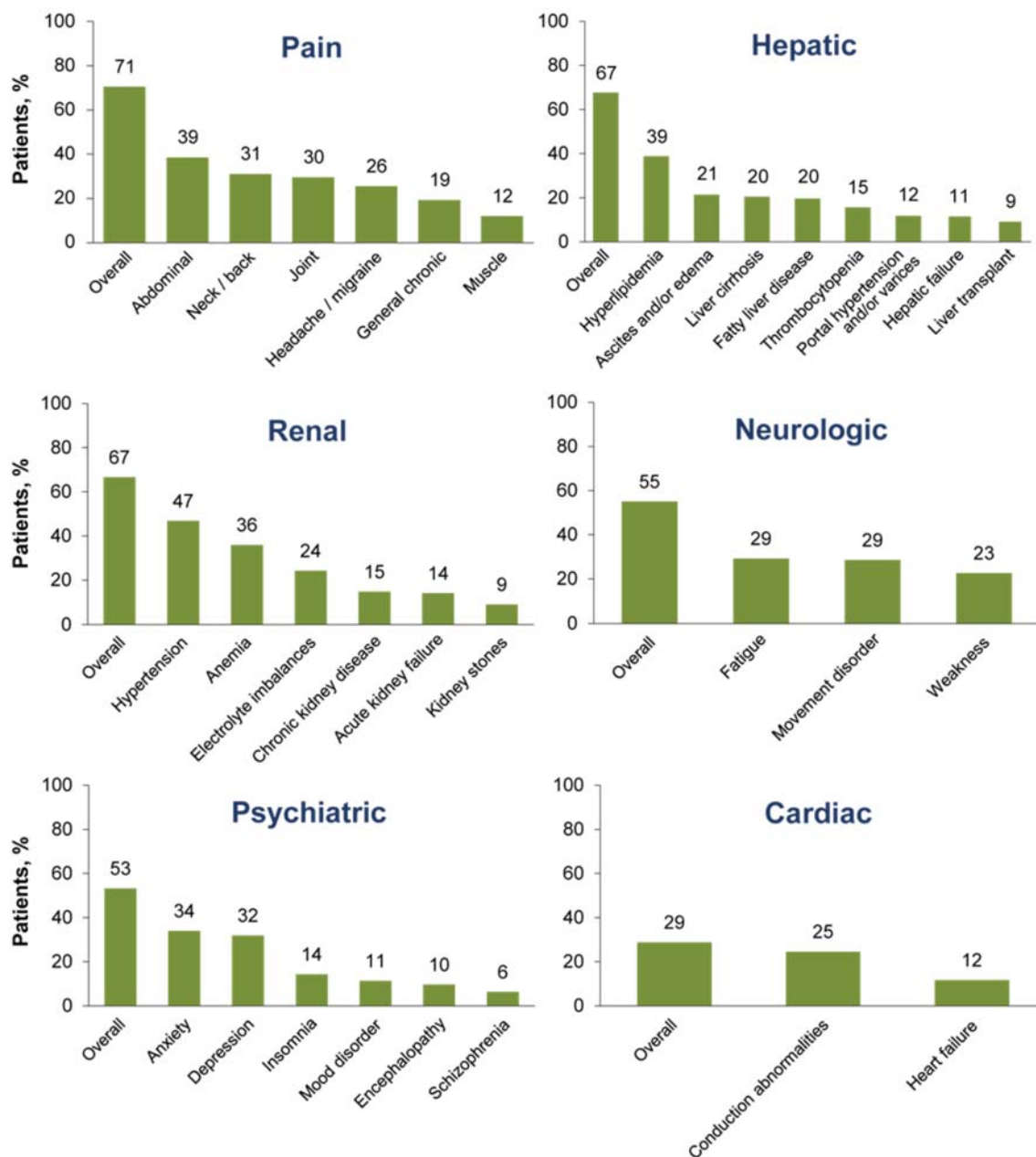
- The greatest proportion of diagnosed complications resulting in clinical care were related to pain, followed by hepatic and renal complications
 - Pain complications most often included abdominal, neck/back, and joint pain
 - Common renal complications included hypertension, anemia, and electrolyte imbalances
- Substantial proportions of patients also had neurologic and psychiatric complications, including fatigue, movement disorders, weakness, anxiety, and depression
- Cardiac complications were less prevalent and commonly included conduction abnormalities and heart failure

Figure 1. Multi-Faceted Complications



RESULTS (CONTINUED)

Figure 2. Summary of Complications Resulting in Care



CONCLUSIONS

- Despite available treatments, Wilson Disease is associated with complications that impact physical and mental health
- Most patients with Wilson Disease received care for at least one hepatic, renal, cardiac, psychiatric, or neurologic complication
 - Pain-related complications were most frequent (71%), followed by hepatic (67%), renal (67%), neurologic (55%), psychiatric (53%), and cardiac (29%)
- This study demonstrates the broad array of complications experienced by patients with Wilson Disease and the unmet need for better treatment options
- This analysis had limitations common to claims data, including the requirement for conditions to be reported in order to be coded, upcoding of complications for claims, and inconsistencies in use of coding systems

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