

Wilson's Disease; Thepaetic Evaluation of Patients Attending Baghdad Teaching Hospital

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Abstract:

Background: *Wilson's disease (WD) is an inherited defectin copper metabolism that causes accumulation of copper in various body organs. It is a potentially curable; if it is diagnosed promptly and treated consistently. Normal life expectancy can be achieved with available treatment.*

Objective: *Therapeutic assessment of a sample of Iraqi patients with WD and finding any association between clinical presentations and studied variables.*

Methods: *A descriptive cross sectional study with analytic elements was conducted during 2011, from the 1st of February till the 10th of June. The sampling method was a convenient non-random one, carried out through consecutive pooling of registered WD patients. A questionnaire-form paper had been developed for the process of data collection.*

Results: *The study had enrolled 29 patients, with a male to female ratio of (1.07:1), their mean age was 27.12 ± 12.18 years. All patients were treated with penicillamine, in addition to dietary modification in 89.7% of them for 18.49 ± 12.51 months. With treatment 69% of patients reported improvement within 2.92 ± 2.48 weeks, 20.7% reported stability, 10.3% deteriorated, therapeutic level of urinary copper was achieved in 23 (79.3%) of patients, serum transaminases levels dropped in 69% and side effects were reported by 58.6% of patients. Death was reported in 3.45% of patients.*

Conclusion:By treating WD patients with D-penicillamine a good therapeutic response; but at least one side effect is reported in most of patients. The best response is in patients with hepato-neurologic type; while those with neuro-psychiatric type have more deterioration and poorer response to treatment compared with other types.

Keywords; wilson, copper, kayser, Therapeutic, Iraq.

Introduction

Wilson's disease (WD) is a rare autosomal-recessive disorder of copper metabolism. It is a hepatolenticular degeneration that characterized by the accumulation of copper in various body organs. It is fatal unless treated, is a potentially treatable condition with the availability of effective pharmacologic therapy, while if not treated; it can cause severe brain damage, liver failure and death[1, 2, 3, 4].

The prevalence of WD is about 1:30,000 and a frequency of heterozygotic carriers is about 1:90. WD resembles 6.9% of chronic liver diseases in Iraq[1,2,3,5,6,7,8].

The availability of effective treatment makes early diagnosis crucial. WD should be considered in any person younger than 30 years with liver disease. The combination of liver disease with extrapyramidal motor abnormalities, or with hemolysis strongly suggests WD and should never be missed[4,9, 10].

Patients with WD have a good prognosis if the disease is diagnosed promptly and treated consistently. The long-term outcome depends on adherence to treatment, without it the disease is fatal, if therapy is started before symptoms develop, symptoms

are rarely seen and prognosis is very good, while many patients die because WD is recognized after appearance of symptoms [1, 7].

By eliminating copper-rich foods from their diet along with an effective lifelong treatment; a normal life-expectancy and an excellent long-term survival for this fatal illness can be offered and most patients improve their liver function within 6–12 months. Three drugs are involved mainly in treatment; D-Penicillamine, Trientine and Zinc salts. In pregnancy treatment must be maintained because any interruption carries a high risk of fulminant liver failure. Liver transplantation is curative as it corrects the metabolic defect[1,2,3,7,9].

Pencillamine is given in a dose of (1.5-2g/day) initially; maintenance (0.75-1g/day, 20mg/kg/day). Monitoring should be done closely initially, and then at least twice a year by clinical examinations, consultation and laboratory tests, including; serum levels of copper and ceruloplasmin and 24-hour urinary copper excretion[1,2,11].

In Iraq WD accounts for 6.9% of chronic liver diseases[8], but; with no available study to assess WD's therapeutic evaluation; thus the objective of this study is to assess therapy regarding; type, duration, response, side effect, death and any association between clinical presentations and other studied variables of a sample of Iraqi patients with WD.

Death was reported in one patient; a 20 yr. old female with neuro-psychiatric presentation, who was 11.5 yr. old at the onset, and after a delay of 30 months she was diagnosed with WD and kept on penicillamine for 6.5yr. till she died; after repeated fits and an advanced functional deterioration. Comparing this with other previous studies Merle et al, 2005; 3 patients died; two because of multiple-organ failure and one because of cholangiocellular carcinoma[12], Medici et al, 2006 death was reported in 3 patients; due to advanced disease and pontinemyelinolysis[13]. Patient with WD is liable to many complications as hemolysis , cirrhosis, fatty liver, hepatitis, infections, bone fractures, injuries, joint

contractures or deformity, muscle atrophy, neuro-psychological complications, loss of ability to care for self or functioning, in addition to side effects and non-compliance. The tragedy is that if a patient stops treatment, probability of death from fulminant liver failure within 1-2 yr. is very high, even if the patient was initially asymptomatic. Liver failure and CNS damage are the most common dangerous effects. If not diagnosed and treated early, WD is fatal[9,11].

Methods

The study is a descriptive cross sectional, with some analytic elements. The data was collected at the Department of Gastroenterology, Baghdad Teaching Hospital of Medical City, Baghdad, Iraq. Data collection was extended from the 1st of February to the 10th of June 2011. The target population was all registered and newly diagnosed WD patients. The sampling method was a convenient non-random one, carried out through consecutive pooling of all WD patients. A data of 29 patients was collected by direct interviews, full clinical assessment and medical records analysis. Patients who accepted to participate in study, completed the required parameters and fit the inclusion criteria during the study period were included in study. The inclusion criteria were; history and clinical examination findings suggestive of WD as: hepatic manifestations, neuropsychiatric manifestations, family history of WD, serum ceruloplasmin level below 200 mg/L[1,2], presence of K-F ring, liver biopsy: if the liver copper concentration more than 250 μ g/g dry weight[1,2], serum copper level below 70 μ g/dl (below 11 μ mol/L)[1,2], 24-hr. urinary copper excretion more than 100 μ g/24-hr.[1,2], and/or positive Penicillamine challenge test (if 24-hr. urinary Copper more than 1600 μ g/24-hr., (more than 25mmol/ L)[1,2]. The patient age should be fulfilling at least three of the above criteria and the last criterion is a definite diagnosis. These diagnostic standards had been proposed basing on Sternlieb's criteria [9, 11].

The exclusion criteria were; age above 60 years, evidence of coexisting liver diseases including; viral hepatitis A, B C or E, chronic liver disease with cholestatic component, Alpha one antitrypsin deficiency, Coomb's positive hemolysis, pregnancy, history of Alcohol intake, history of use of copper containing intrauterine devices (IUCD) or oral contraceptive[5], history of intake of medications that may cause extrapyramidal side effect such as antipsychotic drugs and Metoclopramide and/or history of chorea.

The Statistical Package for the Social Science (SPSS) version 17.0 software had been used for all computerized statistical analysis. Numerical; normally distributed variables was expressed as mean \pm standard deviation; while categorical variables were expressed as frequency, range and percentage. Continuous variables were compared by ANOVA test for the variance analysis and categorical variables were compared by using Chi-square tests. P-value equal or less than 0.05 was considered as a statistically significant.

Results

In the current study, therapeutic evaluation was based on several domains, including; treatment duration, type of treatment, treatment response, side effects and availability of medications.

The patients' total number was 29, males were 15 (51.7%) and females were 14 (48.3%), giving a male to female ratio of (1.07:1). Their mean age was 27.12 ± 12.18 years (yr.) (14-56 yr.). According to their main clinical presentation; patients had been classified into 4 clinical subgroups to facilitate studying them; hepatic in 8 (27.6%) of patients, neuro-psychiatric in 4 (13.8%), mixed hepato-neurologic presentation in 9 (31%), and other presentations in 8 (27.6%).

Therapeutic evaluation of the study sample was based on several domains; duration, type, response, availability of treatment and side effects.

The treatment duration was 18.49 ± 12.51 (ranging between 1-44) months. The treatment was based on D-penicillamine (0.75-1g/day) in all patients in addition to diet and lifestyle modification in 26 (89.7%) of patients, symptomatic treatment in 6 (20.7%) of patients and zinc acetate (150mg/day) in 2 (6.9%) of them initially for two month in addition to penicillamine then continued on penicillamine only.

Concerning subjective response to treatment; 20 (69.0%) of patients reported a subjective improvement within 2.92 ± 2.48 (0.5-8) weeks of treatment, while 6 (20.7%) of them reported a stability in their condition and only 3 (10.3%) of them reported deterioration. Although males reported slightly less (66.7%) improvement than females (71.4%); but males reported more (26.7%) stability than females (14.3%) and less (6.7%) deterioration than females (14.3%) ($P = 0.617$). The 24 hr. urinary copper level during treatment; it was between 200 and 500 $\mu\text{g}/\text{day}$ in 23 (79.3%) of patients, it was seen more in males (93.3%) than females (64.3%) ($P = 0.054$). The drop in the levels of serum transaminases with treatment was seen in 20 (69.0%) of patients, but the response was more in males (80.0%) than females (57.1%) ($P = 0.184$). Death during study period was reported in one (3.45%) patient (that resembles one (7.1%) of female patients) ($P = 0.292$) (Table 1)).

Figure (1) shows that 17 (58.6%) of patients were reported at least a one side effect. Gastric upset was the most frequent side effect, it was reported in 7 (24.1%) of patients, arthralgia was seen in 5 (17.2%) of patients, skin lesions and subjective loss of taste were seen in 3 (10.3%) of patients, for each, neurological worsening and pancytopenia were reported in 2 (6.9%) of patients for each, and proteinuria and febrile reaction were seen in one (3.4%) patient, for each. No patient developed immunological long-term effects (systemic lupus erythematosus, nephritis, positive ANA) as a side effect.

Regarding availability of medications almost all of patients [28(96.6%)] got it from private sector and only one (3.4%) received it from government sector.

Regarding patients' therapeutic evaluation in different clinical subtypes; The longest treatment's duration (36.25 ± 6.18 months) was in those with neuro-psychiatric presentation, while the shortest duration (12.16 ± 11.04 months) was in those with other presentations. Treatment duration was 19.12 ± 13.5 months in patients with hepato-neurologic presentation and it was 16.5 ± 8.45 months in patients with hepatic presentation, and there was statistically significant association between the treatment's duration and clinical types ($P = 0.011$) (Table (2)).

Concerning reported subjective improvement; it was the highest proportion 8 (88.9%) among patients with hepato-neurologic presentation and the lowest 1 (25%) among patients with neuro-psychiatric presentation. Regarding subjective worsening with treatment; it was the highest 1 (25%) among patients with neuro-psychiatric presentation, while there was no reported worsening among patients with pure hepatic presentation. Regarding subjective stability reporting, it was the highest 2 (50.0%) among patients with neuro-psychiatric presentation, and it was not reported in patients with hepato-neurologic presentation ($P = 0.312$) (Table (2)).

The proportion of patients with urinary copper level between (200-500 $\mu\text{g}/24$ hour), as an indicator of good therapeutic response was the highest (100%) among patients with hepato-neurologic presentation and the lowest (25%) among patients with neuro-psychiatric presentation. That was statistically significant ($P = 0.019$) (Table (2)).

The drop in the level of serum transaminases with treatment, which was seen in 20 (69.0%) of participants; it was the highest 8 (88.9%) among patients with hepato-neurologic presentation and the lowest 0 (0%) patients among patients with neuro-psychiatric presentation. That was a statistically significant ($P = 0.007$) "P

value<0.05"). Death during study period was reported in only one (25%) patient of patients with neuro-psychiatric presentation ($P=0.091$ "P value>0.05") (Table (2)).

Regarding medications' induced side effects reported by patients with different clinical presentations; 75% of patients with neuro-psychiatric and with other presentations, for each; 50% of patients with pure hepatic presentation; and 44.4% of patients with hepatoneurologic presentation had developed a one or more medication induced side effects (Table (3) and Figure (2)).

Discussion:

All patients (100%) in current study were treated with D-penicillamine, 89.7% were also managed by diet and life style modification; while symptomatic treatment was required in 20.7%. Treatment was without interruption for 18.49 ± 12.51 months. These results were in agreement with that of other previous studies by; Merle et al, 2005[14]; Panagariya et al, 2006[15]. This can be explained by knowing that choosing D-penicillamine in high percentage of patients because of its approved effectiveness in treating WD. The problem with WD therapy is compliance; as it is hard to convince young people, who feel well, of the need to take medicine 2-4 times daily for the rest of their lives. Patients should eliminate copper-rich foods from their diet in the first year of therapy, after that should take only small amount[1,2,3,7,9,16,17,18].

Regarding current study patients' response to treatment; 69% improved within 0.5-8 weeks of treatment, 20.7% reported stability and 10.3% deteriorated. Males reported more stability than females. Urinary copper level was (200-500 μ g/day) in 79.3%, this was more in males. The drop in serum transaminases was detected in 69%, this was more in males. These results were in agreement with that of (Lowette et al, 2009)[19]; (Panagariya et al, 2006)[15]; (Merle et al, 2005)[12] and (Iorto et al, 2004)[20].

Medications' induced side effects were reported in 58.6% of the patients, gastric upset was the most frequent (24.1%), arthralgia in 17.2% of patients, skin lesions and loss of taste in 10.3%, for each, neurological worsening and pancytopenia in 6.9% for each, and proteinuria and febrile reaction in 3.4%, for each. Comparing results with previous studies; Lowette et al, 2009 reported side effects: 71.4%; dermatological: 43%, and proteinuria: 38% [19], Merle et al, 2005 reported side effects: 74.4%, rash: 15.9%, epigastric discomfort: 1.4%, neurological: 13.8%, arthralgia: 15.9%, proteinuria: 10.9% and pancytopenia: 2.9% [12]. It was expected to report medications' induced side effects; as an early side-effect of penicillamine was an initial neurological worsening in 10-50% of patients. Due to the interference of medication with collagen and elastin formation; skin lesions may be developed. Other side effects vary from minor (loss of taste, gastrointestinal upset, and arthralgia) to severe (proteinuria, leukopenia, or thrombocytopenia), aplastic anemia occurs rarely [1,7,11]. Side effects were higher in previous studies may be because of the fact that the treatment duration in them was much longer than that of this study.

Regarding therapeutic response among patients with different clinical subgroups; patients of hepato-neurologic presentation showed a better response than those of pure hepatic presentation and the last was better than that of neuro-psychiatric presentation, and this difference was significant. Similar results were reported by Merle et al, 2005 [12]. These results can be explained by the fact that; in WD, response and prognosis depend on clinical subtype. And patients with hepatic symptoms are of a better response and prognosis and of shorter delay than those with neurologic symptoms; whose symptoms can persist and worsen despite treatment [1,7,9].

Conclusions

By treating WD patients with D-penicillamine a good therapeutic response can be achieved; but at least one side effect is reported in most of patients. The best response is in patients with hepato-neurologic type; while those with neuro-psychiatric type have more deterioration and poorer response to treatment compared with other types.

Tables and Figures

Table (1): Treatment response of the study group (n=29).

<i>The treatment response indicator</i>		<i>Gender</i>						<i>P value</i>
		<i>Male</i>		<i>Female</i>		<i>Total</i>		
		<i>No</i>	<i>%</i>	<i>No</i>	<i>%</i>	<i>No</i>	<i>%</i>	
Urinary Copper (200-500µg/24 hr.)	Positive	14	93.3	9	64.3	23	79.3	0.054
	Negative	1	6.7	5	35.7	6	20.7	
Drop in transaminases level	Positive	12	80	8	57.1	20	69.0	0.184
	Negative	3	20	6	42.9	9	31.0	
Reported subjective difference	Improvement	10	66.7	10	71.4	20	69.0	0.617
	Worsening	1	6.7	2	14.3	3	10.3	
	Stable	4	26.7	2	14.3	6	20.7	
Death during study period		0	0	1	7.1	1	3.45	0.292
<i>Total</i>		15	100	14	100	29	100	
Duration for subjective improvement[Mean±SD(Range)] week							2.92±2.48(0.5-8.0)	
No; number, %; percent, DF; degree of freedom, P; P value, SD; standard deviation, hr.;hour.								
<i>Note: P value equal or less than 0.05 is considered as a statistically significant; while P value of more than 0.05 is not significant.</i>								

Table (2): Treatment response and treatment duration among patients with different clinical presentations (n=29).

<i>The treatment response indicator</i>	<i>The main initial presentation</i>								<i>P value</i>
	<i>Hepatic</i>		<i>Hepato-neurologic</i>		<i>Neuro-psychiatric</i>		<i>Other</i>		
	<i>No=8</i>	<i>%</i>	<i>No=9</i>	<i>%</i>	<i>No=4</i>	<i>%</i>	<i>No=8</i>	<i>%</i>	
Drop in serum transaminases	7	87.5	8	88.9	0	0	5	62.5	0.007(Sig)
Urinary copper (200-500 µg/24hr.)	7	87.5	9	100	1	25.0	6	75.0	0.019(Sig)
Subjective improvement	6	75.0	8	88.9	1	25.0	5	62.5	0.312
Subjective worsening	0	0	1	11.1	1	25.0	1	12.5	
Subjective stability	2	25.0	0	0	2	50.0	2	25.0	
Death during study period	0	0	0	0	1	25	0	0	0.091
<i>The time duration</i>	<i>Hepatic</i>		<i>Hepato-neurologic</i>		<i>Neuro-psychiatric</i>		<i>Other</i>		<i>P value</i>
	<i>Mean±SD</i>		<i>Mean±SD</i>		<i>Mean±SD</i>		<i>Mean±SD</i>		
Treatment's duration(month)	16.5±8.45		19.12±13.5		36.25±6.18		12.16±11.04		0.011(Sig)
No; number, %; percent, DF; degree of freedom, P; P value, SD; Standard Deviation, hr.;hour, (Sig); Significant.									
<i>Note: P value equal or less than 0.05 is considered as a statistically significant; while P value of more than 0.05 is not significant.</i>									

Table (3): Medications' induced side effects among patients with different clinical presentations (n=29).

The side effect	The main initial presentation								P value
	Hepatic		Hepato-neurologic		Neuro-psychiatric		Other		
	No=8	%	No=9	%	No=4	%	No=8	%	
Any side effect*	4	50.0	4	44.4	3	75.0	6	75.0	0.509
Gastric upset	2	25.0	2	22.2	2	50.0	1	12.5	0.557
Arthralgias	1	12.5	1	11.1	1	25.0	2	25.0	0.833
Skin lesions	1	12.5	1	11.1	0	0	1	12.5	0.908
Subjective loss of taste	0	0	1	11.1	0	0	2	25.0	0.356
Neurological worsening	0	0	0	0	1	25.0	1	12.5	0.297
Pancytopenia	1	12.5	0	0	0	0	1	12.5	0.627
febrile reaction	0	0	0	0	0	0	1	12.5	0.437
Proteinuria	0	0	1	11.1	0	0	0	0	0.512

No; number, %; percent, DF; degree of freedom, P; P value, SD; Standard Deviation
[*The same patient may have more than one side effect].

Note: P value equal or less than 0.05 is considered as a statistically significant; while P value of more than 0.05 is not significant.

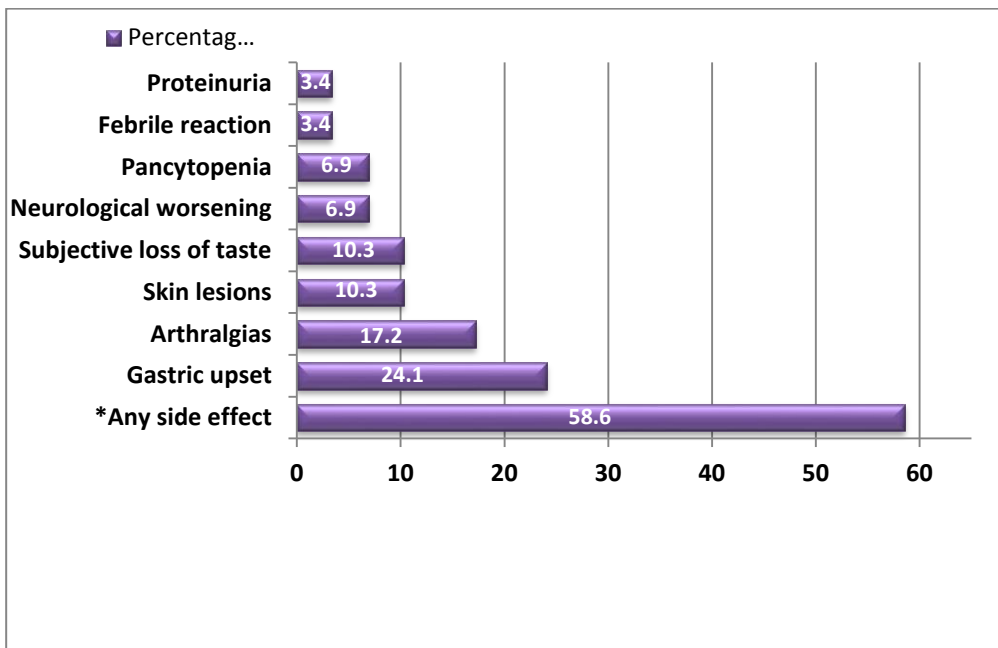


Figure (1): Medications induced side effects in the study group (n=29).

[*The same patient may have more than one side effect].

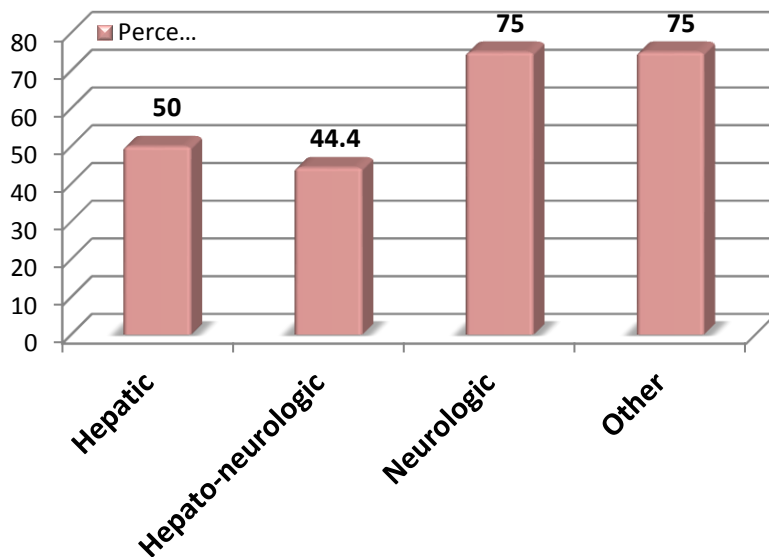


Figure (2): Medications' induced side effects among patients with different clinical presentations [The same patient may have more than one side effect] (n=29).

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التقييم العلاجي للمرضى المصابين بداء ولسون في العراق / مستشفى بغداد التعليمي

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المستخلص:

خلفية البحث: داء ولسون هو خلل وراثي أيضا للنحاس والذي يسبب تراكم النحاس في مختلف اعضاء الجسم. وهو مَرَض قابل للشفاء إذا ما شُخِّصَ وعولج بحزم وبدون تأخير. ويُمكن الوصول إلى العمر المتوقع للمريض مقارنةً بأقرانه من غير المصابين بالمرض بوجود علاج فعال ومتاح.

هدف البحث: التقييم العلاجي لعينة من المصابين بداء ولسون في العراق وايجاد اي علاقة ما بين الانماط السريرية و المتغيرات المدروسة.

طرائق البحث: لقد أُجريت هذه الدراسة الوصفية المقطعية ذات العنصر التحليلي للمدة من الأول من شباط إلى العاشر من حزيران لعام 2011. طريقة أخذ العينات كانت بالاختيار غير العشوائي الملائم المُنفذ بالجمع المتتالي للمرضى المسجلين والمتاحين أثناء فترة الدراسة. جُمع البيانات كان بِمَلاَ استبيان مُعدَّ مُسبقاً و بالمُقابلة المُباشرة مَعَ المَرَضَى.

النتائج: تضمنت الدراسة 29 مريضاً، كانت نسبة الذكور إلى الإناث فيهم (1:1.07). كان متوسط أعمارهم 27.12 ± 12.8 سنة. عولج جميع المرضى بعقار (البنسلامين)، إضافةً إلى الحمية الغذائية في 89.7% منهم لمدّة 18.49 ± 12.51 شهر. وبالمُعَالَجَة أُبْلِغَ عن التحسن في 69% من المرضى خلال 2.51 ± 2.62 اسبوع، استقرار الحالة في 20.7%، تدهورها في 10.3%، تم الوصول الى المستوى العلاجي للنحاس في الادرار في 23 (79.3%) من المرضى، هبطت مستويات أنزيمات الكبد في 69% من المرضى وحدثت آثار جانبية للدواء في 58.6% منهم. سُجِلَت حالات وفاة في 3.45% من المرضى.

الاستنتاجات: بعلاج جميع المرضى المصابين بداء ولسون بعقار (البنسلامين) سجل وجود استجابة علاجية جيدة؛ ولكن سجل على الأقل عرض جانبي واحد لدى اغلبهم. الاستجابة الفضلى لدى المرضى المصابين بالنمط السريري الكبدي العصبي؛ بينما المصابون بالنمط العصبي النفسي فليدهم استجابة علاجية أقل وتدهور أكثر من الباقين.