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High-Value-Care Case Based Management in Pediatric Wilson Disease

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ABSTRACT

High-value care (HVC) is a term used for providing the best care possible for patients while simultaneously decreasing unnecessary expenses. HVC is a good instance of striking a balance between cost and benefit. Principles of value-based health care provide physicians with the necessary knowledge, tools, and strategies to improve the process of clinical reasoning. In this article, we present a case of Wilson's disease in an 8-year-old boy as a clinical example of the impact of HVC on clinical decision-making and clinical reasoning.

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Introduction

Increasing costs of health care is a global concern for patients, governments, and health systems [1]. HVC provides high-quality care for patients while concurrently decreasing unnecessary expenses and the health care system [2]. On the other hand, low-value care (LVC) is defined as performing an intervention that is likely non-beneficial or has minimal benefit. The risk of harm outweighs the benefit, or the extra expenses do not provide extra proportional advantages [3]. A module is designed to ameliorate clinical reasoning skills as a facility to practice HVC by methods such as a bias check-in (accurately collecting information, making several differential diagnoses, considering the worst-case scenario), a diagnostic time-out (consulting with other experts), checklists/ clinical decision support (using electronic resources and articles), and close observation (follow-up of patients for complications) [4]. Comprehending the advantages and disadvantages of an intervention is key to assessing whether it can provide HVC. Comparing strategies or therapies is of importance. Identifying the individuals in whom an intervention is effective, and the differences in effectiveness between subgroups should also be considered [5]. In order to provide HVC, the first step is to reduce or discontinue performing non-beneficial interventions, and the next step is to use effective interventions and reduce costs [5]. Patients, payers, providers, suppliers, and society can benefit from the value-based healthcare system as follows: patients attain better health at a lower cost, providers gain greater patient satisfaction and efficiencies, payers manage costs and reduce risk, suppliers can align their prices with positive patient outcomes, society becomes healthier concurrently overall healthcare costs decrease [6]. It is estimated that more than $10.5 Billion was spent by the commercially insured individuals on the five HVC services in the US in 2015, vaginal deliveries and human immunodeficiency virus therapies contributing the most, while healthy behaviors counseling being the lowest contributor [7]. This article aimed to describe and discuss the management of a case of Wilson's disease based on the principles of HVC. Wilson's disease is an autosomal recessive disorder of copper metabolism. It is caused by a mutation in the ATP7B gene and may affect various organs, including the liver, kidneys, brain, and cornea [8]. The estimated prevalence ranges from 1:29,000 to 1:40,000 [9]. The genetic prevalence is higher due to the incomplete penetrance of the gene or the existence of modifier genes [10]. The initial clinical manifestation is usually between 5 and 45 years [11]. The presentations are mainly hepatic or neurological [12]. Some patients may not present the typical triad of low serum copper, low ceruloplasmin, and high urinary copper levels [10]. Detecting the ATP7B gene is the most efficient method for clinical molecular diagnosis [12]. Asymptomatic hepatic copper deposition in hepatocytes is early involvement. Fatty changes, periportal inflammation, piecemeal necrosis and fibrosis, chronic active hepatitis, and finally, cirrhosis may occur, which can be seen on ultrasound, computed tomography, or magnetic resonance imaging [10]. Lifelong treatment is required even in asymptomatic cases. Treatments for Wilson's disease include D-penicillamine, trientine, and zinc salts [12].

Case Description

An 8-year-old boy was referred to our center complaining about weakness, lethargy, jaundice, tarry stool, and abdominal distension. He was admitted to the pediatric gastroenterology ward after being triaged in the emergency department. On the physical examination, the patient's skin seemed icteric, the conjunctiva was pale, and she had a fever of 100.7 °F. The abdomen was distended. Caput medusae sign was evident. He mentioned going to the hospital and seeing the general practitioner a month before the admission with a complaint of dizziness and tarry stool. The physician justified the symptoms by consuming blackberries and reassured the patient's family. The practitioner had graduated from medical school about one month before (lack of training based on HVC and inexperience). Hospital authorities had just reprimanded him for performing unnecessary laboratory tests. He stated that curiosity and increasing the possibility of diagnosing the disease were the causes of performing the extra tests (LVC). Ever since, he had been cautious about performing laboratory tests, and in some cases, he even sent patients home without a prescription. (Inability to differentiate HVC from LVC). After taking the history and physical examination on admission to our center, the resident physician performed cell blood count and liver function tests, renal function tests, electrolytes tests, an abdominal ultrasound, and a Doppler study.

Table 1: Patient's Laboratory Data

	Patient value	Reference range(male)
WBC	1.3×10^{9}/L	4.4-11.3
RBC	3.32×10^{12}/L	4.50-5.90
HGB	10.1 g/dL	14.0-17.5
HCT	29.4%	41.5-50.4
MCV	88.8 fL	80.0-96.0
MCH	30.3 pg	27.5-33.2
MCHC	34.1%	33.4-35.5
RDW	15.7%	
PLT	29×10^{9}/L	150-450
ALP	169 IU/L	Less than 350
AST	1170 IU/L	10 – 40
ALT	917 IU/L	10 – 40
Albumin	2.1 g/dL	3.4 to 5.4
PT	35 seconds	10-13
PTT	50 seconds	25-36

WBC=white blood cell; RBC=red blood cell; HGB=hemoglobin; HCT=hematocrit; MCV =mean corpuscular volume; MCH =mean corpuscular hemoglobin; MCHC =mean corpuscular hemoglobin concentration; RDW =red cell distribution width; PLT =platelet; ALP=alkaline phosphatase; AST= aspartate aminotransferase; ALT=alanine aminotransferase; PT=prothrombin time; PTT=partial thromboplastin time

A hematology consultation was performed due to the patient's pancytopenia (diagnostic time-out approach). The possibility of cytopenia secondary to portal hypertension was raised. Abdominal and pelvic ultrasounds were performed to investigate liver disease and subsequent hypersplenism. On the ultrasonography, ascites and splenomegaly of 16 mm were reported. Doppler's study revealed hypertension. After evaluating the paraclinical tests (data collection-the first stage of clinical reasoning), the resident physician stated the initial clinical scenario follows: The patient is an 8-year-old boy who presented with the clinical picture of acute liver failure with increased portal blood pressure (making a clinical hypothesis - the second stage of clinical reasoning) so diagnostic tests are needed to find the cause of acute liver failure. Due to the patient's melena and portal hypertension. At first, the patient visited a gastroenterology fellow physician who decided to enlist the patient for esophagogastroduodenoscopy and colonoscopy sessions. At the same time, the gastroenterologist agreed that it was unnecessary to perform a colonoscopy (HVC principal-reduction of an unnecessary procedure). The patient underwent esophagogastroduodenoscopy, and esophageal varices were revealed. After band ligation, diuretics were started to manage the ascites, and the patient went on a salt-restricted diet.

A medical intern examined the patient with a slit lamp and did not find Kayser–Fleischer rings and stated that Wilson's disease might be ruled out. However, the resident physician explained that the Kayser–Fleischer rings might not be seen in all cases of Wilson's disease. Meanwhile, the patient's mother was concerned about a positive family history of hypothyroidism and asked the patient's gastroenterologist to perform thyroid function tests (performing a useless test at the patient's request contrary to the principles of HVC). The gastroenterologist explained that the assessment of thyroid function would not be reliable given her son's current clinical condition, which may cause sick euthyroid syndrome (HVC principle-not performing unnecessary tests). However, the resident physician performed thyroid function tests the next day due to a positive family history recorded in the patient's file (LVC-performing unrequired tests). As expected, the thyroid function tests were not within the normal range, but the patient's mother was reassured that the tests would be carried out again after the patient's full recovery. On the grand round of gastroenterology, the resident physician was asked about the differential diagnoses of the patient. The resident made four differential diagnoses according to the clinical scenario, including drug-induced hepatitis, viral hepatitis, autoimmune hepatitis, and Wilson's disease. Finally, the gastroenterologist made a definite diagnosis (continuing to build a clinical scenario and using bias check-in). The patient mentioned no history of taking medication. So the drug-induced hepatitis was ruled out. A search was conducted using PubMed and Google Scholar databases with the keywords liver failure, children, melena, and pancytopenia, which confirmed the same results of the clinical reasoning. The same was done for the performed tests (clinical decision support tools strategy). Diagnostic tests were done to rule out the remaining diseases (clinical hypothesis test - backward reasoning method). The tests were negative for autoimmune hepatitis and viral hepatitis. Wilson's disease screening test was also negative. A liver biopsy was performed, which did not confirm Wilson's disease (failure to follow the principles of HVC regarding not performing harmful action – owing to the expected levels of urine copper and normal screening tests, it would be better not to perform useless invasive action).

Discussion

A scoring system was suggested at the 8th international meeting on Wilson's disease in Leipzig in 2001, including signs and symptoms and laboratory tests [13]. The total score determines the next steps in approaching patients. In our case, the patient's total score was 2. So the next step was to assess the mutations responsible for Wilson's disease. The patient's family was concerned about the costs of treatment. As Wilson's disease seemed strongly possible, the physician suggested that the tests be performed by the department of the laboratory of a medical school with a 20% reduction in cost (HVC principle - cost-benefit analysis, diagnostic

test with clinical benefit over cost). The cost of genetic testing was estimated and paid for by a charity. The ATP7B gene mutation was discovered. Treatment with D-penicillamine was started, and the patient was given a proper diet and was referred for a liver transplant. In general, for this patient, decisions were made using clinical reasoning, electronic resources, consulting, and financial costs of services. It seems that physicians and health professions students have a poor understanding of the costs of medications and diagnostic tests. Teaching high-value and cost-conscious care to future physicians seems necessary [14]. Educational interventions may help physicians to deliver high-value, cost-conscious health care. Knowledge transmission, reflective practice, and a supportive learning environment are essential factors in the success of educational programs preparing students and physicians for the delivery of HVC [1]. In a study conducted at Johns Hopkins Bayview Medical Center, internal medicine residents participated in two-week rotations to learn the principles of HVC. The sessions included an introduction to the main concepts in HVC, Bayesian thinking, clinical cases, and a review of a patient's hospital bill. Finally, a curriculum was implemented that might ameliorate high-value practice patterns through point-of-care education in medicine wards [15].

Conclusion

This case was presented as a clinical example of the impact of HVC on clinical decision-making and clinical reasoning. As it seems that physicians are not comprehensively familiar with high-value and cost-conscious care, the organization of curriculums to teach HCV principles to residents and medical interns should be taken into consideration.

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