http://ijp.mums.ac.ir
Original Article

Utility of Modeling End-Stage Liver Disease in Children with Chronic Liver Disease

Hamidreza Kianifar [1], Sanaz Jafarzadeh Fakhari[2],*Maryam Khalesi[2], Mohammad Ali Kiani[1], Atefeh Ezzati[3], Seyed Ali Jafari[1]

[1]Department of Pediatric Gastroenterology, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.
[2]Department of Pediatrics, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.
[3]Ghaem Hospital Research Center, Mashhad University of Medical Sciences, Mashhad, Iran.

Abstract

Introduction:
Chronic liver diseases consist of wide spectrum disorders that may be complicated by cirrhosis and therefore need to transplantation. The pediatric end-stage liver disease (PELD) score and model of endstage liver disease (MELD) score has been used as predictors of mortality chronic liver diseases listed for liver transplantation. The aim of this study is evaluation of relation between PELD\MELD score and evidence of cirrhosis in children with choronic liver disease.

Materials and Method:
This cross-sectional study conducted on 106 patients of chronic liver disease referred to Ghaem haspital, Mashhad university of medical science, Iran during 24 months period (2010-2013). PELD and MELD score were calculated for all patients. Clincal and patholoogical findings of cirrhosis were recorded.

Results:
Mean age of patients was 68/3 ± 41.8 months. Mean PELD\MELD score was -1/59± 9/64. There was significant correlation between PELD\MELD score and clinical icter, spelenomegaly, evidence of hepatopulminary syndrome, esophageal varices, evidence of cirrhosis in tissue specimences.

Conclusion:
PELD\MELD score appear to be benefit for detection of cirrhotic children among paients with choronic liver disease.

Keywords:
Choronic Liver Disease, Cirrhosis, PELD\MELD Score.

Introduction

Several prognostic parameters have been advocated for children with chronic liver diseases. The pediatric end-stage liver disease (PELD) model and model of endstage liver disease (MELD) are new models for scoring liver disease severity. PELD scores can be calculated using 5 parameters for patients less than 12 years: age, growth retardation, serum bilirubin, serum albumin and INR. MELD scores also can be calculated using 3 parameters

***Correspondence Author:**
Assistant Professor of Pediatrics, Ghaem Medical Center, Mashhad University of Medical Sciences, Mashhad, Iran.
E-mail: khalesim@mums.ac.ir
Received date: Nov 23, 2013
Accepted date: Dec 10, 2013

for patients younger than 12 years: serum bilirubin, serum creatinine and INR(1-6). In this study we evaluated the validity of the PELD risk scoring system as a severity index for diagnosis of cirhosis in chronic liver disease, which is a major indication for liver transplantation in children.

Materials and Methods

This cross sectional study was carried out among all cosequtive children younger than 18 years with chronic liver disease referred to Ghaem haspital, Mashhad university of medical science, Iran during 24 months period (2010-2013). Patient with liver malignancy, concurrent hemolytic disease or protein loosing enteropathy were excluded. Demographic data, clinical signs and symptoms of cirhosis and paraclinical findings were recorded in a checklist for all patients. PELD and MELD score were calculated with PELD\MELD calculator for patients younger and older than 12 years old retrospectively. After entering data into SPSS software version 16.0, association between demographic, clinical, and laboratory data were analyzed by x^2 or Fisher exact test for quantitative data and by t test or Mann-Whitney U test for qualitative data. P value <0.05 was considered significant.

Results

Between December 2010 and November 2013, 106 patients with chronic liver disease were evaluated. Mean age of patients was 68.3 ± 41.8 months. Mean PELD\MELD score was -1.59 ± 9.64 with range of -16-26. Demoghraphic, clinical, and PELD\MELD findings of the patients are summarized in Table1. Five variables had significant correlation with PELD\ MELD score including clinical icter, spelenomegaly, evidence of hepato-pulminary syndrome, esophageal varices, evidence of cirrhosis in tissue specimences (Table.1).

Table1: Demoghraphic, clinical, and PELD\ MELD findings of the patients

	No (%)	PELD\MELD score (mean ± SD)	P value
Sex(Female)	56(52.8)	-	-
Age,mean(range),month	68.3 ± 41.8(12-156)	-	-
Failure to trive	55(51.9)	-	-
Icter	47(44.3)	3.44 ± 9.21	0.0001
clubbing	21(19.8)	1.71 ± 7.09	0.07
hepatomegaly	78(73.6)	-1.19 ± 8.98	0.47
Splenomegaly	47(44.3)	1.25 ± 9.63	0.06
Encephalopathy	4(3.8)	-2.0 ± 0.0	0.66
Ascites	15(14.2)	0.73 ± 8.44	0.27
HPS*	8(7.5)	10.5 ± 8.84	0.004
Esophageal varices	13(12.3)	5.38 ± 12.45	0.005
Cirrhosis**	9(8.5)	10.66 ± 7.9	0.0001

*hepatopulmonary syndrome, **pathological evidence of cirrhosi

Discussion

Prognostic models are useful to estimate disease severity and survival as well as to make decisions regarding specific medical interventions. These models are developed using analytical methods that determine the effects on specific outcomes such as death of variables of interest eg, demographic data and laboratory values.

PELD score has been proposed as an objective tool to prioritize children awaiting orthotopic liver trans- plantation (OLT);

a higher PELD score has been associated with increased pre-OLT mortality (7). Previous studies showed it was also associated with a higher rate of complications and mortality as reported in previous studies (7,8).

Our finding showed there were significant relation between presence of icter, spelenomegaly, HPS, encephalopathy, esophageal varices and evidence of cirrhosis in biopsy specience and PELD\MELD score. These parameters are clinical and pathological evidence of cirrhosis in chronic liver disease. Also there was no significant relation between hepatomegaly and PELD\MELD score. It may be due to shirinkage of cirrhotic liver. Also there was no significant relation between clubbing and encephalopathy and PELD\MELD score. It may be due to low incidence of clubbing and encephalopathy in chronic liver disease in camparision to other findings of cirrhosis.

Jagadisan etal evaluated clinical presentation, etiology, outcome, and mortality in children with an acute hepatic insult superimposed on chronic liver disease. Children with acute on chronic liver disease (ACLD) Were divided into two groups: acute on chronic liver failure (ACLF) and non-ACLF. 17 patients with ACLF criteria and 19 with non-ACLF were inrolld. The 3-month mortality of ACLF group was significantly higher than non-ACLF group (59% vs 11%, P¼0.001). PELD score of >25.5 predicted death, with sensitivity of 100% and specificity of 83.3% (9). Several studies have shown that PELD/MELD profiling is a useful scoring system that can be utilized for selecting patients with end stage liver disease(2,4,10). On the other hand some studies bring up some limitation for these scoring system, such as the effect of age, gender, and low body mass in chronic liver disease on serum creatinine level that may introduce a bias independent from severity of liver disease. For evaluation PELD\MELD score validity we sugesst more studies with larger sample size and evaluation of labratory liver function tests.

Conclusion

It seems that evaluation of PELD\MELD score can be a useful method for screening cirrhotic children among paients with choronic liver disease.

Acknowledgment

The authors thank all children and parents with CLD. We would like to thank reasearch development center of Ghaem Hospital for their assistant in this manuscript. This study was supported by a grant from the vice chancellor for reasearch of mashhad univercity of medical sciences. This manuscript is part of student thesis with approval nomber of 6939.

References

1. Shinkai M, Ohhama Y, Take H, Fukuzato Y, Fujita S, Nishi T. Evaluation of the PELD risk score as a severity index of biliary atresia. Journal of pediatric surgery. 2003 Jul; 38(7): 1001-4. PubMed PMID: 12861526.

2. Wiesner RH, McDiarmid SV, Kamath PS, Edwards EB, Malinchoc M, Kremers WK, et al. MELD and PELD: application of survival models to liver allocation. Liver transplantation : official publication of the American Association for the Study of Liver Diseases and the International Liver Transplantation Society. 2001 Jul;7(7):567-80. PubMed PMID: 11460223.

3. Freeman RB, Jr., Wiesner RH, Roberts JP, McDiarmid S, Dykstra DM, Merion RM. Improving liver allocation: MELD and PELD. American journal of transplantation : official journal of the American Society of Transplantation and the American Society of Transplant Surgeons. 2004;4 Suppl 9:114-31. PubMed PMID: 15113360.

4. Barshes NR, Lee TC, Udell IW, O'Mahoney CA, Karpen SJ, Carter BA, et al. The pediatric end-stage liver disease (PELD) model as a predictor of survival benefit and posttransplant survival in pediatric liver transplant recipients. Liver Transpl. 2006 Mar;12(3):475-80.

5. McDiarmid SV, Anand R, Lindblad AS, Principal I, Institutions of the Studies of Pediatric Liver Transplantation Research G. Development of a pediatric end-stage liver disease score to predict poor outcome in children awaiting liver transplantation. Transplantation. 2002 Jul 27;74(2):173-81.
6. Sanchez MC, D'Agostino DE. Pediatric end-stage liver disease score in acute liver failure to assess poor prognosis. J Pediatr Gastroenterol Nutr. 2012 Feb;54(2):193-6.
5. McDiarmid SV, Anand R, Lindblad AS, Principal I, Institutions of the Studies of Pediatric Liver Transplantation Research G. Development of a pediatric end-stage liver disease score to predict poor outcome in children awaiting liver transplantation. Transplantation. 2002 Jul 27;74(2):173-81.
6. Sanchez MC, D'Agostino DE. Pediatric end-stage liver disease score in acute liver failure to assess poor prognosis. J Pediatr Gastroenterol Nutr. 2012 Feb;54(2):193-6.
7. Dehghani SM, Gholami S, Bahador A, Haghighat M, Imanieh MH, Nikeghbalian S, et al. Comparison of Child-Turcotte-Pugh and pediatric end-stage liver disease scoring systems to predict morbidity and mortality of children awaiting liver transplantation. Transplant Proc. 2007 Dec;39(10):3175-7.
8. McDiarmid SV, Merion RM, Dykstra DM, Harper AM. Selection of pediatric candidates under the PELD system. Liver Transpl. 2004 Oct;10(10 Suppl 2):S23-30.
9. Jagadisan B, Srivastava A, Yachha SK, Poddar U. Acute on chronic liver disease in children from the developing world: recognition and prognosis. J Pediatr Gastroenterol Nutr. 2012 Jan;54(1):77-82.
10. Freeman RB Jr, Gish RG, Harper A, Davis GL, Vierling J, Lieblein L, et al. Model for end-stage liver disease (MELD) exception guidelines: results and recommendations from the MELD Exception Study Group and Conference (MESSAGE) for the approval of patients who need liver transplantation with diseases not considered by the standard MELD formula. Liver Transpl. 2006 Dec;12(12 Suppl 3):S128-36.