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The Plasma Amino Acid Profile and its Relationships to Standard Quantities of Liver Function in Infants and Children with Extrahepatic Biliary Atresia and Preterminal Liver Cirrhosis

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Summary: The absolute and relative concentrations of 16 plasma amino acids in 48 mostly dystrophic infants and children (median of age 1 ½ years) with extrahepatic biliary atresia and mainly stable preterminal cirrhosis were compared with those of controls. Patient plasma amino acid data were analysed statistically for diagnostic usefulness and correlated with standard biochemical quantities of liver function and of liver perfusion. In the patients the total amounts of non-essential and essential amino acids were reduced by 19%, and with the same significance ($p < 0.0005$). Plasma tyrosine was increased (+40%), while taurine (−44%) and branched chain amino acids (+28.8% to −34.7%) were decreased. Methionine values varied widely. In the molar fractional plasma amino acid profile, only alanine, valine, and leucine were decreased, while threonine, methionine, tyrosine, phenylalanine, ornithine, and serine were increased. Discriminate function analysis showed that the plasma amino acid data discriminated 93.8% of the patients from controls. The concentrations of some amino acids in plasma seemed to have been influenced by protein-calorie deficiency in the patients. The valine/tyrosine ratio and the *Fischer* index (ratio branched chain/aromatic amino acids) were significantly reduced in the patients versus controls (1.54 ± 0.55 vs 3.08 ± 0.55 and 1.66 ± 0.39 vs 3.00 ± 0.48). A number of significant correlations (range of r: 0.37 − 0.59, $p < 0.05$, 30–48 data pairs) were calculated between plasma amino acid data and several standard biochemical quantities of liver function. The statistical analyses also showed that the *Fischer* index began to decrease gradually and linearly early in the progression of liver failure. It is concluded that plasma amino acid data can be useful in the evaluation of the progression of liver failure and possibly of the nutritional status in liver transplant candidates with biliary atresia.

Introduction

Extrahepatic biliary atresia is a congenital structural defect of the hepatic biliary tree. Its frequency is estimated to be 1 : 8000 to 1 : 15000. It is responsible for 75 to 80% of the cases of conjugated hyperbilirubinaemia in children (1, 2). In the natural course of extrahepatic biliary atresia, 70 to 90% of cases die within 2–3 years (3). Affected patients may benefit, for a time, from hepatoportoenterostomy (4), but most eventually develop biliary cirrhosis and progressive liver failure (5, 6). The only definitive therapy appears to be liver transplantation (3, 6). Lack of donor liver and low body weight in recipients are serious problems, so that staging and optimal nutritional care of the chronically malnourished transplantation candidates are vital (1, 3). Single laboratory findings will usually not be indicative of impending terminal liver failure in extrahepatic biliary atresia (3).

In adult cirrhotics, plasma concentrations of the aromatic amino acids are increased and those of branched chain amino acids are decreased. Methionine can be markedly elevated (7–9). The *Fischer* index (the ratio of the sum of the concentrations of