INTRODUCTION: Dietary treatment with a phenylalanine (phe) restricted diet from newborn period and long life sustained is successfully in PKU. However, adherence to strict diet decrease over the years. Some PKU patients respond to pharmacological doses of tetrahydrobiopterin (BH4). Different tests have been developed to select the responsive patients. Our main experience is with short-term 48 hs test. A correct interpretation of these tests, need an increase of blood phe level between 6-8 mg/dl through diet. Patients and family concern and professional disregard of actual phe tolerance, allow misinterpreting test results. Objectives Prove that an individual plan of feeding with fixed amounts of phe and menus which was previously agreed with family and patient allows more reliable test results, standardized diet Maintained all the test long. Recalculate phe tolerance of patients.

Methods and material An observational and retrospective study was performed. 29 patients were tested with BH4, using the 48-h test according Blau. Tests were divided in 2 groups. Group A composed by 18 patients from 2011-2013 with no standardized diet previous test. Group B composed by 11 patients from 2014 to 2018 with standardized diet. Phe tolerance is defined as the amount of phe (mg/day) that maintains blood phe concentrations within the target range. Tolerance was compared between the estimated previous test and after six months of BH4 tests. Statistical analysis was performed with SPSS Statistics 18. From each patient, the coefficient of percentage variation was calculated to compare the variability of the Phe dosages performed on the initial day. The differences between groups were evaluated by a Mann-Whitney test. RESULTS: Group B showed a minor variability, statistically significative (p-value=0.043) in relation to day 0 of the test (without BH4). 72% of patients were over restricted before test and showed a higher tolerance after the test, even in patients continuing with diet only. CONCLUSIONS: Standardized diet allowed a minor variability of basal levels of phe, which is essential for assessing impact of BH4 on phe levels. Time and difficulties of reaching adequate phe values prior test was caused by over-restriction of diet.