**INTRODUCTION:** PKU patients need individual follow up performed by a metabolic team. **OBJECTIVE:** to describe the management strategy of PKU patients in our clinic. **PATIENTS AND METHODS:** 133 patients (2 months to adulthood) from different Argentinian provinces and other Latin American countries (Ecuador, Bolivia and Paraguay) are followed up by a metabolic team (two doctors, one dietitian and one neuropsychologist) that works close to the laboratory where blood samples are processed. **RESULTS:** After diagnosis, patients come to consultation monthly during the first year of age and then every four months. Phenylalanine (Phe) levels are tested fortnightly until age five and monthly thereafter. According to Phe tolerance current patients were classified as severe (n: 41) moderate (n: 39); mild (n: 9) and hyperphenylalaninemic (n: 44). Thirteen patients were late diagnosed (7 from Ecuador). A neurocognitive evaluation is performed yearly or at least before starting kindergarten, elementary school, high school and college. Treatment is instituted individually, considering clinical, social and emotional needs in every patient. 72 patients receive conventional treatment: diet + protein substitute which is indicated as amino acids mixture (n: 66), in two patients combined with glycomacropeptides (GMP) in 2 only as GMP. Forty-two patients receive just diet counseling. Six patients (5 late diagnosed) are treated with neutral aminoacids combined with protein substitute and diet. 37 patients underwent a dihidrochloride of saprotptereine (BH4) test. 4 were tested as neonates while 33 were tested later with a 48 hours test (n: 28) or with a long term test (n: 5). 31 patients were responders (decrease from basal Phe blood level ≥30%). Thirteen of them began treatment with BH4. Four were able to withdraw protein substitute and nine continue with it: six as amino acid mixture and three combining also GMP. 5 women (1 moderate PKU, 1 mild PKU and 3 hyperphenylalaninemas) underwent successful pregnancies. Both PKU received conventional treatment and hyperphenilalaninemas diet counseling. With acceptable adherence, our early detected PKU patients grow and develop normally. Late detected patients have improved their quality of life. **CONCLUSION:** Our team works with international recommendations but individual assessment of affected children is provided and treatment designed accordingly.