The aim of this dissertation is to present a summary of today's knowledge about the maternal phenylketonuria syndrom and to compare it with the 15-year experience from the Metabolic Centre of the Faculty Hospital Kralovské Vinohrady. The studied group consisted of 29 women, who gave birth to 41 children (22 of them originated from a pregnancy with a phenylalanin-restricted diet introduced before conception). The observed antropometric features of these children (height, weight, head circumference), which were measured after birth as well as in the first year of the life, show statistic significantly lower values in comparison with the national average. Totally 7 cases of a minor damage to the children were reported. Among 4 others some damage in a form of an inborn heart defect was observed. The occurance of the pathological effects was connected with high levels of phenylalanin in the maternal blood and its frequency was nine times higher among pregnancies without the diet. The intrauterine exposition of the child to phenylalanin showed a very significant negative correlation with the head circumference of the child (meassured after the birth and at the age of one year), as well as with the height in the first year. The best outcomes of a pregnancy may be expected when an optimal maternal metabolic compensation with levels of maternal phenylalanin less than 6 mg/dL is achieved before conception and continued throughout the pregnancy.