Succinylacetone (SA, 4, 6 diketoheptanoic acid) is primary diagnostic metabolite for inherited disorder hepatorenal tyrosinemia type I [1]. Hereditary tyrosinemia type I is an autosomal recessive disorder caused by deficiency in fumarylacetoacetate hydrolase (FAH) in the tyrosine degradation. The deficiency of FAH leads to an accumulation of the metabolites fumarylacetoacetate and maleylacetoacetate, which together form succinylacetone. Since succinylacetone is not found in normal body fluids, the presence of succinylacetone in urine, blood or amniotic fluid is pathognomonic of tyrosinemia [2].

This work describes the capillary electrophoretic determination of SA which examined by means of capillary zone electrophoresis. All electrophoretic measurements were performed with an Agilent capillary electrophoresis system equipped with a diode-array detector. The data processing was carried out with the Agilent Chemstation software. The detection wavelength was set to 300 nm. The separation was performed with reversed polarity mode at -28kV voltage. Injections were made at 5.10^3 MPa for 10s. The fused silica capillary used for experiments had 50μm inner diameter. The total length of capillary was 56cm. The fused silica capillary was conditioned prior to use by rinsing in sequence with 1mol/L NaOH for 20 min, 0.1mol/L NaOH for 20 min, water for 5 min, and then buffer solution for 10 min. The capillary was washed with 1mol/L NaOH for 5 min, 0.1mol/L NaOH for 5 min, water for 3 min, and buffer for 10 min in between runs. Borate and phosphate buffer solutions with different pHs and with various additives such as cetyltrimethylammonium bromide (CTAB) and diethylenetriamine (DETA) were used and compared for SA detection. Best results were obtained when using a phosphate buffer (50mmol/L, pH 9.6) which was containing 20mmol/L NaCl and 0.05mmol/L CTAB as the cationic surfactant in a fused silica capillary.

The developed method seems promising due to its simplicity and reliability for SA determination in biological samples.

References