Altered calpain caspase cascade in CF cells
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Cystic Fibrosis (CF) is a common, lethal, autosomal recessive disease caused by mutations in the gene encoding the cystic fibrosis transmembrane conductance regulator (CFTR). In the most frequent mutation (F508del), F508del-CFTR protein is misfolded and retained in the endoplasmic reticulum (ER). We previously showed that the unfolded protein response (UPR) may be triggered in CF. Since prolonged UPR activation leads to apoptosis via the calcium-calpain-caspase-12-caspase-3 cascade and because apoptosis is altered in CF, our aim was to compare the ER stress-induced apoptosis pathway between wild type (WT) and F508del-CFTR expressing cells. Using cells endogenously expressing the mutated CFTR and their corrected counterparts we show that the calpain-caspase cascade is altered in F508del-CFTR expressing cells. We propose that this alteration is involved in the altered apoptosis triggering observed in CF.

The activity of components of macromolecular complex cathepsin C–cathepsin A–neuraminidase–beta-galactosidase in mixed saliva of cystic fibrosis patients
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Introduction: Cystic fibrosis (CF) is a complex disease with multiple clinical symptoms. The clinical picture of the disease can’t be only explained by the malfunction of CFTR various enzymatic disturbances has been described in CF patients. Some proteolytic and glycolytic enzymes are at least partly involved in pathogenesis of chronic Pseudomonas aeruginosa (PA) infection in CF patients.

Aim of the study: In our study we assessed the activity of cathepsin C (CatC), cathepsin A (CatA), neuraminidase (Neu), beta-galactosidase (Gal) in mixed saliva of 66 CF patients (CF) and 66 healthy controls (HC).

Methods: We assessed total concentration of sialic acid in saliva, correlation of studied parameters with chronic PA infection, the age of acquiring of chronic infection. The Receiver Operating Characteristic (ROC) curve analysis was also performed for assessed sialometric parameters.

Results: The activity of CatC and Gal was significantly higher in CF group, the activity of CatA was significantly lower in CF group and activity of Neu was comparable in CF group and HC. Total sialic acid was significantly higher in CF. There was no correlation between the activity of studied enzymes and chronic PA infection, but in ROC analysis the activity of CatC and Gal differentiate CF from HC.

Conclusions: The activity of components of macromolecular complex in mixed saliva of CF patients is higher comparing to HC. Despite the poor correlation with PA infection, the enzyme activity can be useful as additional diagnostic test in CF patients.