The aim of the project is to search for the mechanism involved in the formation of the genetic mosaicism which occurs in women carriers of mutations in three generations. In Polish family affected with Barth syndrome we identified a pathogenic mutation c.83T>A, p.Val28Glu in the TAZ gene (Zapała B and Płatek T, Annals of Human Genetics, February 2015). Mutation was identified in 2 boys with clinical signs of the syndrome and in 7 women mutation carriers. In 5 of them mutation occurred in a mosaic form- the mutation was absent in the blood while it was present in the epithelial cells. Only 2 women present mutation both in the blood and epithelial cells while one of them had only a small percentage mutation in the blood and full heterozygous mutation in the epithelial cells. These results suggest inheritance of genetic mosaicism. The project aims to investigate whether the occurrence of genetic mosaicism, shown by several generations of the family, are due to processes associated with cytogenetic rearrangements. On the example of described family in our publication and other patients-carriers' of other mutations in the gene *TAZ* we want to examine whether tafazzin could disrupt meiotic and mitotic cells divisions. The project is expected to study changes in the form of cytogenetic balanced/ unbalanced chromosome aberrations and the study of maternal uniparental disomy. These studies may explain if the presented mosaicism is only a point mutation or comprises a larger fragment of the X chromosome, and possibly also relates translocations, deletions, duplications or inversions which involve other chromosomes. These results may indirectly indicate if tafazzin within a pathogenic mutation was identified is involved in the regulation of cell division. As has been demonstrated in a mouse model lack of tafazzin impairs meiotic cell divisions at the stage of pachytene what was shown by Cadalbert and co-workers in 2015 (Cadalbert et al. 2015). To examine that we wanted to perform cytogenetic analysis of microarrays type CGH + SNP and by the classical karyotyping method in all persons involved in the project. Karyotyping using the proposed arrays allows for the simultaneous detection of gene copy number variants, deletions and duplications, and the loss of heterozygosity (LOH) and uniparental disomy (UPD). These studies are reasonably likely to explain the presence of genetic mosaicism, which is inherited in the presented family.To conclude the project can confirm and explain the mechanism of transmission of genetic mosaicism to offspring. Perhaps the project will help to clarify unequivocally whether in the described family and other patients there are additional genetic disturbances in the form of chromosomal aberrations that could confirm the study of Cadalbert and colleagues that tafazzin can control cell division. The results of the project may be important for the development of the field because for the first time it is present a case showing the phenomenon of inherited mosaicism in 3 generational family, which also remains under a big sign an inquiry about the mechanism of inheritance and compliance with the rules of Mendelian inheritance. Studies on the example the described family seem to be a very good model to study this type of disorders.