Ts1Cje mouse model for Down syndrome research

ABSTRACT

Intellectual disabilities, hypotonia and cranio-facial dysmorphism are the cardinal characteristics of Down syndrome (DS) individuals. To varying extent, DS individuals also exhibit other developmental problems such as heart defects, vision impairments, hearing loss, hypothyroidism, dental problems and gastrointestinal defects. They are also at a higher risk for certain disorders such as early onset neurodegeneration and childhood leukaemia. For many decades, scientists have been trying to elucidate how additional full or partial set of chromosome 21 may responsible for these developmental disabilities or disorders. To date, many investigations are based on molecular, cellular and behavioural analyses of mouse models exhibiting similar characteristics observed in DS individuals. Among various models, Ts1Cje, in particular, is suitable for dissecting the effect of additional genetic materials on learning and memory impairment as well as muscle weakness in DS. Ts1Cje has partial triplication of the mouse chromosome 16, which is syntenic to chromosome 21 in human. This talk will focus on the genetics of Ts1Cje mouse model for DS and discuss how much do we know about the model and the degree of resemblance between Ts1Cje and human DS individuals in term of neuropathology of Down syndrome.