Geneeskundige Stichting Koningin Elisabeth
Fondation Médicale Reine Elisabeth
Königin-Elisabeth-Stiftung für Medizin

Wetenschappelijke prijzen
Prix scientifiques
2014

Prof. dr. Claudia Bagni (KU Leuven)
Dr. Fadel Tissir (UCL)
Dr. Laurent Nguyen & dr. Brigitte Malgrange (ULg)
Prof. dr. Vincent Timmerman, PhD & prof. dr. Peter De Jonghe, MD, PhD (UA)
Prof. dr. Pierre Maquet & dr. Christophe Phillips, ir (ULg)

Wetenschappelijke prijzen/Prix scientifiques 2014

UCB Award 2014
€ 100.000
Overhandiging van de/Remise du "UCB Award"
H.K.H. Prinses Astrid/S.A.R. la Princesse Astrid & monsieur Ismail Kola

Laureaat/Lauréat:
Prof. dr. Claudia Bagni (KU Leuven)
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Onderzoeksproject/Projet de recherche:
mRNA metabolism at synapses and spine remodeling: Insights into Fragile X, Autism and Schizophrenia.

Memory formation and cognitive processes rely on activity-dependent synaptic plasticity. Synapses are specialized structures of the spines, the functional protrusions of neuronal cells required for connectivity, integration and brain functions.
Dendritic spines are known to change shape and to appear and disappear entirely in physiological and pathological conditions.
It has long been hypothesized that such changes may be the basis of memory itself. Synaptic inputs dictate the time, place and amount of protein synthesis necessary for correct neuronal functions and the understanding of how spines are continuously shaped during our life remains one of the most exciting and important questions in neuroscience.
Dysregulation of cellular and molecular mechanisms regulating neuronal maturation leads to spine dysmorphogenesis and to a variety of pathological conditions including the most common form of inherited mental disability, the Fragile X Syndrome (FXS), due to the absence or mutation of a single protein, the Fragile X Mental Retardation Protein (FMRP).
We had previously shown that FMRP, together with its cytoplasmic interactor CYFIP1, controls in an activity dependent manner, the synthesis of key proteins at synapses. The protein CYFIP1 has a critical role in human brain function/s. Deletions and/or duplications of CYFIP1 have been associated to several neurological disorders.
We have now shown that CYFIP1 is involved in the formation of synaptic processes and that unbalances in CYFIP1 interactive networks result in neuronal dysfunctions such as dendritic spines dysgenesis. Furthermore, the identified CYFIP1 interactome opened new perspectives to define regulatory pathways shared by neurological disabilities, characterized by dysregulated CYFIP1 expression, such as Fragile X Syndrome, Autism, Schizophrenia and Alzheimer Disease.

Wetenschappelijke prijzen/Prix scientifiques 2014

Prijs/Prix Burggravin/Vicomtesse Valine de Spoelberch
€ 100.000
Overhandiging van de prijs/Remise du prix
H.K.H. Prinses Astrid/S.A.R. la Princesse Astrid & madame Eric Speeckaert

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Onderzoeksproject/Projet de recherche:
Celsr genes in brain development and function.

Le développement du système nerveux est un long processus qui, chez l'humain, commence dès la 3ème semaine de gestation et ne s'achève qu'après la puberté. Il assure la mise en place des réseaux neuronaux fonctionnels de l'adulte. Rien dans ce processus n'est laissé au hasard. Au contraire, toutes les étapes font l'objet d'un contrôle génétique très strict dont le moindre dysfonctionnement conduit à des conséquences dévastatrices. Il est donc essentiel d'étudier les mécanismes moléculaires du développement cérébral car leur compréhension aiderait à terme à développer des stratégies thérapeutiques en cas de pathologies et/ou de lésions du système nerveux.
Afin d'approcher le développement cérébral, nous utilisons des modèles murins que nous développons au laboratoire. Au cours des dix dernières années, nos efforts se sont focalisés sur une famille de trois protéines (dites CELSRs) impliquées dans la polarité cellulaire. Au début des années 2000, deux membres de la famille (CELSR1 et CELSR2) étaient répertoriées dans les bases de données, mais rien ou presque n'était connu quant à leurs fonctions. Nous avons eu la chance d'identifier le troisième membre (CELSR3). Nous avons montré que les trois protéines sont abondamment exprimées dans le système nerveux où elles jouent des rôles cruciaux dans la prolifération des cellules souches, la migration neuronale, le câblage du système nerveux, et l'homéostasie cérébrale.

Wetenschappelijke prijzen/Prix scientifiques 2014

Solvay Prize
€ 25.000
Overhandiging van de/Remise du "Solvay Prize"
H.K.H. Prinses Astrid/S.A.R. la Princesse Astrid & Jonkheer Jacques van Rijckevorsel

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Onderzoeksproject/Projet de recherche:
Unravelling the roles of lysine acetylation in neural development.

In humans, primary cilia are found on nearly every cell in the body. By acting as a "sensor", these extensions have emerged as a key structure in a broad array of homeostatic and developmental processes. Although this immotile organelle has been discovered decades ago, little is known about its biology but accumulating evidence shows that mutations in cilia genes can lead to malformations underlying neurological or psychiatric disorders, as well as deafness and balance disorders.
Recent studies suggest that protein acetylation contributes to brain development and the goal of the present project is to decipher if and how this process controls primary ciliogenesis. We recently discovered that the Elongator complex promotes protein acetylation in the developing cerebral cortex. In addition, our preliminary results showed that Elongator is localized in the primary cilium of brain progenitors and in its absence the length of cochlear cilia is modified. Therefore, we will use a conditional knockout mouse line available in our laboratory to impair the Elongator complex activity in the developing brain and the inner ear. By combining a multidisciplinary approach, we will identify the proteins targeted by Elongator whose acetylation is required for primary ciliogenesis. Our work will contribute to shed some new light on new mechanisms that control the development and homeostasis of the cerebral cortex and the inner ear in health and disease.

Wetenschappelijke prijzen/Prix scientifiques 2014

Prijs/Prix Baron van Gysel de Meise
€ 12.500
Overhandiging van de prijs/Remise du prix
H.K.H. Prinses Astrid/S.A.R. la Princesse Astrid & le Baron van Gysel de Meise

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Onderzoeksproject/Projet de recherche:
De ziekte van Charcot-Marie-Tooth (CMT): van gen en proteïne netwerken tot ziekte mechanismen.

CMT is een erfelijke aandoening van het perifeer zenuwstelsel. Er zijn 80 genen beschreven waarin mutaties CMT of een gerelateerde neuropathie veroorzaken. Een aantal genen hebben een specifieke functie in de perifere zenuw, echter voor de meeste is hun functie onduidelijk. Het GSKE project heeft bijgedragen tot het vinden van mutaties in 4 nieuwe genen. Het leverde een belangrijke bijdrage tot de diagnose van CMT bij kinderen. Daarnaast vonden we interacties tussen mutant 'small heat shock' proteïne, tubuline en neurofilament; relevant voor het transportsysteem in zenuwcellen. Tijdens dit project schreven we 26 publicaties en behaalden 4 onderzoekers hun doctoraat.

Wetenschappelijke prijzen/Prix scientifiques 2014

Prijs/Prix Janine et Jacques Delruelle
€ 12.500
Overhandiging van de prijs/Remise du prix
H.K.H. Prinses Astrid/S.A.R. la Princesse Astrid & Baron et Barones Delruelle

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Onderzoeksproject/Projet de recherche:
Characterization of human sleep/Wake regulation using multimodal functional imaging in populations stratified on the polymorphism of PERIOD3 gene.

At any point in time, wakefulness, and its associated cognitive functions, is regulated by the interplay between two processes. First, sleep homeostasis which keeps track of time awake and accumulates sleep need during wakefulness, while sleep need dissipates during sleep. Then, the circadian timing system which counters sleep need during the day to maintain wakefulness, and promotes sleep at night to maintain sleep. At the macroscopic level of the brain function, the interplay between these processes is fairly well characterized, notably through studies we conducted with the support of the FMRE/GSKE. At the microscopic level, the brain mechanisms sustaining this interplay remain poorly understood.
In our research project, we used a novel technique coupling the EEG to transcranial magnetic stimulation (TMS) to assess neuronal function during a normal waking day and following sleep deprivation. Our analyses show that the circadian timing system and sleep homeostasis have an important impact on the responsiveness (the excitability) of the neurons of the cortex. This novel finding constitutes key information on how wakefulness and cognition are regulated during. These results could bear important implication for shift-work, accident prevention and cognition optimization.