

DD/SP du CS

23 Mars 2004

1/AO AFM/INSERM CELLULES SOUCHES

BUCKINGHAM Margaret, Dr labo BUCKINGHAM, Paris, France

Projet: The myogenic, endothelial and hematopoietic potential of stem cells in the dorsal aorta and the extension of this study to adult vessels

LEMARCHAND Patricia, Dr labo ESCANDE, Nantes, France

Projet: Cardiac cell therapy

MOULY Vincent, Dr labo BUTLER-BROWNE, Paris, France

Projet: Myogenic potential of stem cells

RIPOCHE Jean-marie, Dr labo MOREAU, Bordeaux, France

Projet: Cell therapy of tissue fibrosis with mesenchymal stem cells genetically modified to overexpress the gene of matrix metalloproteinase-1

2/ APPEL D'OFFRES DEVELOPPEMENT DES THERAPEUTIQUES

Groupe E: Thérapies génique et cellulaire

Aides aux études

ABDERRAHIM-FERKOUNE Anissa, Dr labo AILHAUD, Nice, France

Projet: Transdifferenciation des preadipocytes en cellules de muscle lisse

ANDRE Franck, Dr labo PERRICAUDET, Villejuif, France

Projet: concepts and mechanisms of the in vivo DNA electrotransfer in muscles

BALDESCHI Christine, Dr labo ORTONNE, Nice, France

Projet: Preclinical gene therapy of dystrophic epidermolysis bullosa

BENRAISS Abdellatif, Dr labo BOITARD, Paris, France

Projet: Gene therapy for metachromatic leucodystrophy based on intracerebral gene transfer using AAV vector

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BERTOLDI Didier, Dr labo GUICHENEY, Paris, France

Projet: NMR imaging of hamster cardiomyopathy: longitudinal study and evaluation of IGF-1 gene therapy

BREJOT Thomas, Dr labo HEARD, Paris, France

Projet: Spinal cord injury repair

BRISSAULT Blandine, Dr labo AUVRAY, Evry, France

Projet : Structure-activity studies of triblocs copolymers. Application to gene transfer in muscle

BRIZARD Mara, Dr labo MALLET, Paris, France

Projet: Gene therapy of ALS: improvement of GDNF delivery by gutless vectors and by controlling gene expression in a mouse model

CHEN Fengtian, Dr labo HEARD, Paris, France

Projet: Investigation of transport and delivery of the lysosomal enzymes IDUA and NaGlu in the central nervous system

DIOP Dialo, Dr labo DENOULET, Paris, France

Projet : Stop codon recognition and readthrough induction in human cells: implications for the treatment of genetic disease.

FRALEY Andrew, Dr labo BEHR, Illkirch, France

Projet: Gene repair mediated by oligonucleotides

GARINOT Marie, Dr labo SCHERMAN, Paris, France

Projet : Ciblage et vectorisation chimique d'ADN pour la thérapie génique : synthese et étude physico-chimique de nouveaux polymeres pour l'administration systématique des complexes ADN/vecteurs

HASSANI Zahra, Dr labo DEMENEIX, Paris, France

Projet: Targeted delivery of RNAi into the neural stem cells of the mouse brain

JACQUIN-BECKER Clarisse, Dr labo MARTIN, Strasbourg, France

Projet : Suppression of a pathological nonsense mutation in human mtDNA by import of engineered cytoplasmic suppressor tRNAs

KIM Félix jackson, Dr labo SITBON, Montpellier, France

Projet : Structure modulaire de l'enveloppe HTLV, motifs clés pour la reconnaissance du récepteur, identification du récepteur et implications en thérapie génique

LAVENU-BOMBLED Cécile, Dr labo ROMEO, Paris, France

Projet: Targeting proteins to storage granules: a new gene-cell therapy strategy



MARODON Gilles, Dr labo BLANCHARD, Montpellier, France

Projet: Preclinical risk evaluation for hematopoietic stem gene therapy: model of ZAP-70 deficiency

MASSOUDI/RODRIGUEZ Ali, Dr labo DANI, Nice, France

Projet: Muscle regeneration by novel human adult multipotent stem cells

MATECKI Stefan, Dr labo PETROF, Montréal, Quebec, Canada

Projet: Strategies for therapy of respiratory muscle failure in muscular dystrophy.

MESEL/LIU Mariana, Dr labo KLATZMAN, Paris, France

Projet : Heart transplantation tolerance induction by gene therapy involving a conditional TK suicide gene.

MONTIER Tristan, Dr labo FEREC, Brest, France

Projet: Development and optimisation of ex vivo and in vivo approaches for gene therapy using synthetic transfection agents

PONS Bénédicte, Dr labo BEHR, Illkirch-Graffenstaden, France

Projet: Correction génique par des oligonuclétides modifés

PRIVATO Marta, Dr labo MOULLIER, Nantes, France

Projet : Gene therapy of Crigler-Najjar disease via in vivo retrovirus-mediated gene transfer to the liver

SIMON Delphine, Dr labo MANDEL, Illkirch, France

Projet: Gene therapy development for Friedreich ataxia: evaluation of efficacy of an echocardiographic directed plasmid injection approach

SKANDER Nadia, Dr labo KREMER, Montpellier, France

Projet: Developing safe, effective and long-term therapy for neurolodegeneration in LSD using canine adenovirus vectors

SOUAMES/GILBERT Sémi, Dr labo BUCHETON, Montpellier, France

Projet: Design of site-specific vectors derived from non-LTR retrotransposons for gene therapy

STEINBERG Marcos, Dr labo JEANTEUR, Montpellier, France

Projet: Preclinical gene therapy studies for ZAP-70 deficiency

STEPHAN Lionel, Dr labo TREMBLAY, Sainte-Foy, Québec, Canada

Projet : A tolerance induction protocol for myoblast transplantation

TRAJCEVSKI Stéphane, Dr labo KLATZMANN, Paris, France



Projet: Replicative and semi-replicative retroviral vectors for cancer gene therapy

TSITOURA Eliza, Dr labo EPSTEIN, Villeurbanne, France

Projet: Development of herpes simplex virus type 1 (HSV-1)-derived vectors for long term and regulatable expression in the brain

Subventions

ANTIGNAC Corinne, Dr labo ANTIGNAC, Paris, France

Projet: In vitro and in vivo gene transfer studies for cystinosis

ARSENIJEVIC Yvan, Dr labo ARSENIJEVIC, Lausanne, Suisse

Projet: Human retinal stem cells for transplantation studies in animal models of retinal degeneration: preclinical studies

BEHR Jean-paul, Dr labo BEHR, Illkirch, France

Projet: Gene repair mediated by oligonucleotides

BODAGHI Bahram, Dr labo KLATZMANN, Paris, France

Projet: Application for the development of an inovative clonotypic model of autoimmune uveitis

BOULANGER Pierre, Dr labo BOULANGER, Lyon, France

Projet: Targeting Adenoviral (Ad) vectors: Ad with restricted tropism and selective internalization into target cells

BUCHETON Alain, Dr labo MECHALI, Montpellier, France

Projet: Design of site-specific vectors derived from non-LTR retrotransposons for gene therapy

CARLE Georges, Dr labo GAUDRAY, Nice, France

Projet: Towards cell therapy for infantile malignant osteopetrosis: using the OC/OC mouse as a model system

CARNAC Gilles, Dr labo MANGEAT, Montpellier France

Projet: Characterization of molecular machanisms underlying determination and differentiation of satellite cells in order to improve muscle repair and regeneration

CARTIER-LACAVE Nathalie, Dr labo BOITARD, Paris, France

Projet: Gene therapy for metachromatic leukodystrophy based on intracerebral gene transfer using AAV vector

CHERADAME Hervé, Dr labo AUVRAY, Evry, France



Projet: Structure-activity studies of triblocs copolymers. Application to gene transfer

in muscle

COSSET François-loic, Dr labo COSSET, Lyon, France

Projet: Upgrading of lentiviral vector for gene delivery to muscle cells in vivo

COSSU/BLOT Giulio/Stéphane, Dr labo COTTARD, Milan, Italie

Projet: Cell therapy of muscular dystrophy by intra-arterial delivery of donor or autologous, genetically corrected mesoangioblasts in the dystrophic dog

COULOMBEL Laure, Dr labo PESCHANSKI, Créteil, France

Projet: Modulation of stem cell fate by short exposure to purified Notch ligands;: potential therapeutic interest

DANI Christian, Dr labo AILHAUD, Nice, France

Projet: Muscle regeneration by novel human adult multipotent stem cells

DAVIES Kay E., Dr labo DAVIES, Oxford, UK

Projet: Studies towards the upregulation of utrophin in DMD

DE VERNEUIL Hubert, Dr labo DE VERNEUIL, Bordeaux, France

Projet: Development of new lentiviral vectors with a selection system for the gene therapy of hematological diseases.

DEGLON Nicole, Dr labo AEBISCHER, Lausanne, Suisse

Projet : Altering the transcriptional program as potential therapeutic approach for Huntington's disease

DEMENEIX Barbara Dr labo DEMENEIX, Paris, France

Projet: Targeted delivery of RNAi into the neural stem cells of the adult mammalian brain

DICKSON George, Dr labo DICKSON, Londres, UK

Projet: Therapeutic inhibition of exon splicing in dystrophin pre-mRNA by specific antisense oligo ribonucleo tides (splicomers): studies in the GRMD canine model of DMD.

DICKSON George, Dr labo DICKSON, London - UK

Projet: Development of AAV vectors encoding human and canine microdystrophin cDNAs for preclinical DMD gene therapy studies.

EPSTEIN Alberto-luis, Dr labo COUBLE, Villeubanne, France

Projet: Development of HSV-1-derived helper-free amplicons for strong and stable transgenic expression.



FEREC Claude, Dr labo FEREC, Brest, France

Projet: Development and optimization of ex vivo and in vivo approaches for gene therapy using synthetics transfection agents.

FERRY/MOULLIER Nicolas/Philippe, Dr labo FERRY, Nantes, France

Projet: Gene therapy for liver inherited diseases using viral vectors: applications to Crigler-Najjar disease

FICHELSON Serge, Dr labo ROMEA, Paris, France

Projet: Transdifferentiation of human hematopoietic stem cells in an early embryonic environment

FROMES Yves, Dr labo GUICHENEY, Paris, France

Projet: Preclinical evaluation of IGF-1 gene transfer into the heart muscle for symptomatic treatment of dilated cardiomyopathies.

GACHE Yannick, Dr labo ORTONNE, Nice, France

Projet: Gene therapy of DEB: Preclinical studies of canine recessive DEB (RDEB) and development of ex vivo gene therapy for correction of dominant DEB (DDEB).

GHERARDI Romain, Dr labo GHERARDI, Creteil, France

Projet: Stem cell therapy of genetic dilated cardiomyopathies

HEARD Jean-michel, Dr labo HEARD, Paris, France

Projet: Gene therapy for neurodegeneration in Hurler and Sanfilippo syndromes

JEAN-JEAN Olivier, Dr labo DENOULET, Paris, France

Projet : Stop codon recognition and readthrough induction in human cells: implications for the treatment of genetic disease

KARPATI George, Dr labo KARPATI, Montréal, Quebec, Canada

Projet: Molecular therapies for dystrophin deficiency

KREMER Eric j., Dr labo KREMER, Montpellier, France

Projet : CAV-2 receptors, trafficking and vectorology : feasibility of therapy for neurodegenerative disorders

LETURCO France, Dr labo DELPECH, Paris, France

Projet: A new strategy of allele-specific vectorized targeted exon skipping for transcriptional correction therapy of selected cases of dystrophinopathy (Duchenne Muscular Dystrophy).

MALLET Jacques, Dr labo MALLET, Paris, France



Projet: Development of lentivirus vectors for regulated expression of therapeutic genes in the central nervous system

MALLET/MURA Jacques/Henri, Dr labo MALLET, Paris, France

Projet: Experimental gene therapy for Retinitis Pigmentosa: lentiviral transfer of genes of the GDNF family in the rd10 mouse and the RCS rat.

MENASCHE Philippe, Dr labo CARPENTIER, Paris, France

Projet: Development of dual-marker system for quantitative studies of myoblast survival and proliferation after transplantation, and improvement of these parameters

MESTAS Jean-louis, Dr labo CHAPELON, Lyon, France

Projet: Ultrasound-mediated Gene therapy: improving the efficacy of transfection through cavitation monitoring

MIDOUX Patrick, Dr labo BELOEIL, Orléans, France

Projet: Vascular portal for gene delivery: dynamic studies of the transendothelial transport of polymer-based gene delivery systems

MIR Luis, Dr labo PERRICAUDET, Villejuif, France

Projet: Concepts and mechanisms of the in vivo DNA electrotransfer in muscles.

MOULLIER Philippe, Dr labo MOULLIER, Nantes, France

Projet: Viral vectors and in vivo application in non human primates demande faite sur 1 an

MOULY Vincent, Dr labo BUTLER-BROWNE, Paris, France

Projet: Injection of human satellite cells: optimisation of a model for cell mediated gene therapy

NEHRBASS Ulf, Dr labo NEHRBASS, Paris, France

Projet: Analyzing intranuclear routing and targeted integration of HIV-based vectors

PAYEN Emmanuel, Dr labo LEBOULCH, Paris, France

Projet: Oncogenic safety Lentiviral Vectors and Gene correction

PELEGRIN-ZURILLA Mireia, Dr labo BLANCHARD, Montpellier, France

Projet: Antibody-based gene/cell therapy of crohn disease

PERRICAUDET Michel, Dr labo PERRICAUDET, Villejuif, France

Projet: Vectors towards TGFBeta receptor

PITARD Bruno, Dr labo ESCANDE, Nantes, France



Projet: A novel class of polymeric synthetic vectors promotes high gene expression in skeletal and cardiac muscles

PRATS Anne-catherine, Dr labo PRATS, Toulouse, France

Projet: Identification of muscle specific IRES (intenal Ribosome Entry Site) to develop a new regulatable vector: a biotechnological tool for gene therapy

PUCCIO Hélène, Dr labo MANDEL, Illkirch, France

Projet: Gene therapy development for Friedreich ataxia: evaluation of efficacy of an echocardiographic directed plasmid injection approach

PUYMIRAT Jack, Dr labo PUYMIRAT, Sainte-Foy, Québec, Canada

Projet: RNA-based methods as potential gene therapy for myotonic dystrophy

ROLLING/MOULLIER Fabienne/Philippe, Dr labo MOULLIER, Nantes, France

Projet: transfer in the retina

SALVETTI/MOULLIER Anna/Philippe, Dr labo MOULLIER Nantes, France

Projet: Factors influencing replication and assembly of the Adeno-Associated Virus (AAV) and its derived recombinant vectors

SIMONELIG Martine, Dr labo MECHALI, Montpellier, France

Projet: Drosophila model for the study and therapy of human oculopharyngeal muscular dystrophy (OPMD)

SKUK Daniel, Dr labo TREMBLAY, Ste Foy, Québec, Canada

Projet: Improving the control of acute rejection aigu for myoblast transplantation: pre-clinical study in non-human priates

SOUDAIS Claire, Dr labo CASANOVA, Paris, France

Projet: Gene therapy for a novel hereditary disease impairing anti-mycobacterial immunity: the complete lack of expression of the interferon gamma receptor

TAYLOR/KLATZMANN/NORAZ Naomi/David/Nelly, Dr labo BLANCHARD, Montpellier, France

Projet: Preclinical risk evaluation for hematopoietic stem cell gene therapy: Model of ZAP-70 deficiency

TORRENTE Yvan, Dr labo BRESOLIN, Milan, Italie

Projet: Ex vivo expansion of Human Muscle-Derived Stem Cell from normal and dystrophic tissues

TREMBLAY Jacques, Dr labo PUYMIRAT, Quebec, Canada



Projet: Improvement of muscle regeneration following myoblast transplantation by blocking myostatin

VILLARD Jean, Dr labo REITH, Genève, Suisse

Projet: Cellular and gene therapy for MHC class II deficiency

WATT Diana, Dr labo WATT, Brighton, UK

Projet: Probing the role of galectin-1 in muscle determination and differentiation in vivo

WELLS Dominic, Dr labo WELLS, London, UK

Projet: Functional assessment of gene therapy for duchenne muscular dystrophy.

YAFFE/NUDEL David/Uri, Dr labo YAFFE, Rehovot, Israel

Projet : A subpopulation of myogenic cells isolated from skeletal muscle Đ possible application for cell therapy

Groupe F : Développement des Thérapeutiques

Aides aux études

FISSON Sylvain, Dr labo KLATZMANN, Paris, France

Projet: Specific tolerance in autoimmune disease using CD4+CD25+ regulatory T-cells: a new approach for the treatment of Polymyositis

GUARDIOLA Philippe, Dr labo GLUCKMAN, Paris, France

Projet: The effects of cellular stress on human FANC deficients cells using a genomic/proteomic approach

OLLIVIER Karen, Dr labo HOGREL, Paris, France

Projet: Physiological response to muscular exercise of patients with McARDLE's disease. Effects of an endurance training programme

SABIL Abdelkebir, Dr labo DEMONGEOT, La Tronche, France

Projet: Optimization of mechanical ventilation in patients with neuromuscular disease

SCHLEMMER Dimitri, Dr labo MUNNICH, Paris, France

Projet: Coenzyme Q10 in energy deficiency in children: incidence, mechanisms and molecular bases, therapeutic implications

SPEER Oliver, Dr labo WERNER, Zurich, Suisse



Projet: Energy metabolism in Duchenne dystrophic muscle and neurons: a potential target for pharmacological interventions

VOISIN Vincent, Dr labo BAUX, Gif sur Yvette, France

Projet: Arginine derivatives for the treatment of Duchenne Muscular Dystrophy

Subventions

ALLAMAND/ROUSSET Valérie/Jean-Pierre, Dr labo GUICHENEY/JACQUET, Paris, France

Projet : Antibiotic-mediated readthrough of stop codon mutations responsible for Muscular Dystrophies

BASTIN Jean, Dr labo MUNNICH, Paris, France

Projet : Pharmacological correction of Carnitine Palmitoyle Transferase 2 (CPT2) deficiency by agonists of Peroxisome Proliferator Activated Receptor (PPAR)

BLOT Stéphane, Dr labo BLOT, Maisons Alfort, France

Projet: Evaluation of clinical and histological effects of a glucocorticoid derivative, NCX-1015, in GRMD dogs.

DE LA PORTE Sabine, Dr labo BAUX, Gif sur Ycette, France

Projet : Arginine derivatives and NO donnors for the treatment of Duchenne Muscular Dystrophy

FOUGEROUSSE Françoise, Dr labo DANOS, EVRY France

Projet: Evaluation of functional and histological effects of a glucocorticoid derivative, NCX 1015, in mdx mouse.

GLUCKMAN Eliane, Dr labo GLUCKMAN, Paris, France

Projet: Gene therapy of sickle cell disease and B-thalassemia

HERON/EYMARD Delphine/Bruno, Dr labo EYMARD, Paris, France

Projet: Sleep evaluation in the childhood type of myotonic dystrophy

LAFORET Pascal, Dr labo EYMARD, Paris, France

Projet : Epidemiologic study and natural history of patients with late-onset acid maltase deficiency in Francence : a preliminary to therapeutic trials

LOPEZ DE MUNAIN Adolfo, Dr labo LOPEZ DE MUNAIN, San Sebastian, Espagne

Projet : A multi parametric approach to natural history knowledge of calpainopathies for efficacy assessment of therapies



MEININGER Vincent, Dr labo MEININGER, Paris, France

Projet: Efficacy, tolerability and safety of minocycline in patients treated with amyotrophic lateral sclerosis (ALS) receiving riluzole

PESCHANSKI Marc, Dr labo PESCHANSKI, Créteil, France

Projet: Thérapie cellulaire et génique appliquée à la maladie de Hungtington, étude clinique et expérimentale

RUSTIN/ROTIG Pierre/Agnès, Dr labo MUNNICH, Paris, France

Projet : Q10 in energy deficiency in children : incidence, mechanisms and molecular bases, therapeutic implications

SEGALAT Laurent, Dr labo COUBLE, Villeurbanne, France

Projet : C. elegans as a tool to screen for compounds slowing dystrophin-dependent muscle degeneration

TZARTOS Socrates, Dr labo TZARTOS, Athènes, France

Projet : Development of a specific therapy for myasthenia gravis by the selective ex vivo withdrawal of patients' anti-AChR antibodies

WALLIMANN Theo, Dr labo WERNER, Zurich, Suisse

Projet: Energy metabolism in Duchenne dystrophic muscle and neurons: a potential target for pharmacological interventions.

3/ APPEL D'OFFRES DECRYPTHON

LAPORTE/MANDEL Jocelyn/Jean-Louis, Dr labo MANDEL, Illkirch, France

Projet: Characterization of the myotubularin homologues and prediction of protein domains and tissue-specific functions

POCH Olivier, Dr labo MANDEL, Illkirch, France

Projet: Identification and analysis of therapeutic targets in pathogenic bacteria implicated in human diarrhoea

PUJOL/MANDEL Aurora/Jean-Louis, Dr labo MANDEL, Illkirch, France

Projet: Insight into the molecular basis in X-adrenoleukodystrophy: Comparative genomics of the peroxisomal proteome and related fatty acid metabolic pathways

ZINN-JUSTIN Sophie, Dr labo Menez, Gif-sur-Yvette, France

Projet: Using the Decrypthon database for the identification of domains in proteins involved in the genome protection



4/ HORS APPEL D'OFFRES

Aides aux études

BENSAMOUN Sabine, Dr labo GOUBEL, Compiègne, France

Projet : Caractérisation multi-échelles (de l'organe à la molécule) des propriétés mécaniques et morphologiques du tissu musculo squelettique (os et muscle)

BOURGEOIS Patrice, Dr labo FONTES, Marseille, France

Projet: Development of therapeutial approaches for CMT 1A

CALBO Sébastien, Dr labo TRON, Rouen, France

Projet : Ovalbumin skeletal muscle transgenic mice as a preclinical model of immunoregulatory therapy of inflammatory myopathies

CHEVESSIER Frédéric, Dr labo GUICHENEY, Paris France

Projet: Characterization of tubular aggregates in skeletal muscle

ESTRADA Francisco-Xavier, Dr labo SAHEL, Paris, France

Projet : Identification et caractérisation du complexe sarcoglycan-sarcospan dans la rétine de souris sauvages et KO Dp71

FARGE Géraldine, Dr labo ALZIARI, Aubière, France

Projet : Caractérisation d'un modèle animal présentant des altérations de l'ADN mitochondrial similaires à celles observées dans les pathologies humaines.

FEDERICI Thais, Dr labo NETO, Rio de Janeiro, Brésil

Projet: Mechanisms of adenoviral vectors entry into neural cells

HADDAD Hafedh, Dr labo GUICHENEY, Paris, France

Projet : Effet de la L-arginine sur l'expression de l'utrophine

KOCAEFE Cetin, Dr laboDANOS, Evry, France

Projet : Mécanismes moléculaires sous-tendant la transformation graisseuse du muscle dystrophique.

LAVANYA Madakasira, Dr labo SITBON, Montpellier, France

Projet : Mise au point de nouveaux outils d'enveloppes rétrovirales pour un adressage préférentiel aux tissus nerveux et musculaires

LEMONNIER Jérôme, Dr labo MELKI, Evry, France

Projet : Etude du rôle de SMN dans les processus de fusion membranaire et cellulaire



LOUFRANI Laurent, Dr labo HENRION, Paris, France

Projet : Role of the cytoskeletal protein dystrophin in the mechanotransduction of pressure and flow in vascular cells

NEGRE Olivier, Dr labo LEBOULCH, Paris, France

Projet: Oncogenic safety Lentiviral Vectors and Gene correction

PERRIN Valérie, Dr labo DEGLON, Lausanne, Suisse

Projet : Altering the transcriptional program as potential therapeutic approach for Huntington's disease

TAVEAU Mathieu, Dr labo DANOS, Evry, France

Projet: Contributions of the study of calpain 3's function and biology

Subventions

ARSEP, Président BRUNEL, Paris, France

Financement de deux *Projet*s sur la sclérose en plaques, par l'intermédiaire de l'ARSEP.

BERARD Carole, Dr labo BERARD, Pierre-Bénite, France

Projet : Complément de financement pour la validation de l'échelle d'évaluation des traitements pour les maladies neuromusculaires.

CENTRE DE BIOTHERAPIE, Boulogne-Billancourt, France

Projet : Construction d'un Centre de biothérapie dans le Groupe Hospitalier Pitié Salpétrière

CHU NANTES, Dr labo DRENO, Nantes, France

Projet : Production de produits cellulaires dérivés de la thérapie cellulaire et génique du CHU de Nantes

CUTIS LAXA, Association CUTIS LAXA

Projet : campagne de prélèvements

DREYFUSS Gideon, Dr labo DREYFUSS, Philadelphie, USA

Projet: The molecular functions of SMN and a search for therapeutics for spinal muscular atrophy.

ENVA/BLOT Stéphane, Dr labo BLOT, Maisons Alfort, France

Projet : Fonctionnement de l'UETM (Unité d'Etude et de Traitement des Myopathies Canines) pour 2003.



ENVN/Nantes, Nantes, France

Projet: Fonctionnement du Centre de Boisbonne pour 2003.

FERREIROAnna, Dr labo Guicheney, Paris, France

Projet: Early-onset myopathies with core lesions; from clinics to therapy through molecular and cell biology. Financement d'un salaire dans le cadre de l'appel d'offres AVENIR (Inserm)

FREZAL Jean, Dr labo FREZAL, Paris, France

Projet : Base de données GENATLAS

GARCHON Henri-Jean, Dr labo GARCHON, Paris, France

Projet: Genetic susceptibility to autoimmune myasthenia gravis: role of the HLA complex and of the genes coding the target autoantigen, the muscle acetylcholine receptor

GOOSSENS Michel, Dr labo GOOSSENS, Créteil, France

Projet: Application to purchase a DNA chip-based molecular workstation

GVPN/BOSCH Fatima, Dr labo BELLETERRA, Barcelone, Espagne

Projet: Gene vector production network

GVPN/TRONO Didier, Dr labo TRONO, Genève, Suisse

Projet: Gene vector production network

GVPN/MOULLIER Philippe, Dr labo MOULLIER, Nantes, France

Projet: Gene vector production network

HEARD Jean-Michel, Dr labo HEARD, Paris, France

Projet: Gene therapy for neurodegeneration in Hurler and Syndrome sanfilippo

IRME, Président FOURTY Dominique, Paris, France

Financement de trois *Projet*s sélectionnés par le Conseil Scientifique de l'IRME dans la thématique « Thérapie génique et cellulaire »

KAMOUN/MUNNICH Pierre/Arnold, Dr labo MUNNICH, Paris, France

Projet de plate-forme de spectrographie de masse à la disposition des équipes de recherche soutenues par l'AFM

KLATZMANN David, Dr labo KLATZMANN, Paris, France

Projet: Traitement des maladies auto-immunes compliquées, d'une myélo-dysplasie/leucémie secondaire par allogreffe gèno-identique de cellules souches hématopoïétiques et de lymphocytes T du donneur génétiquement modifiés exprimant le gène HSV-TK



LAFORET Pascal, Dr labo EYMARD, Paris, France

Projet : Etude de l'efficacité et de la tolérance du salbutamol dans la dystrophie musculaire FSH.

LAOUDJ Dalila, Dr labo BENNES, Montpellier, France

Projet : Analyse protéomique de la dystrophie Facio Scapulo Humerale (landouzy-dejerine)

LE GUERN Eric, Dr labo AGID, Paris, France

Projet: Molecular and cellular study of autosomal recessive forms of Charcot-Marie-Tooth

LEGAY Claire, Dr labo LEGAY, Paris, France

Projet: Biologie des jonctions neuromusculaires normales et pathologiques

MALISSEN Bernard, Dr labo MALISSEN, Marseille, France

Projet : Accélérateur d'inactivation génique AFM-CIML

MAIRE Pascal, Dr labo DAEGELEN, Paris, France

Projet : Demande de soutien pour les voyages et séjours des participants aux réunions du « Club Myologie » organisées par Pascal Maire.

MANDEL/LAPORTE Jean-Louis/Jocelyn, Dr labo MANDEL, Illkirch, France

Projet: Functional studies of the protein implicated in myotubular myopathy and creation of animal and cellular models.

MARCELLE/JAGLA Christophe/Krzysztof, Dr labo DASTUGUE, Clermont-Ferrand, France

Projet: Identification of novel genes regulating proliferation and renewal of satellite cells.

PAQUIS Véronique, Dr labo PAQUIS, Nice, France

Projet : Demande de co-financement pour l'achat d'un appareil à DHPLC

PAULIN Denise, Dr labo PAULIN, Paris, France

Projet: Rôle du facteur SRF (Serum Response factor) dans la régulation des gènes impliqués dans l'activité transcriptionnelle des différents types de muscles : aspects structuraux, dynamiques et thermodynamiques des complexes formés avec SRE - Accueil du Dr Josef STEPANEK

PESCHANSKI Marc, Dr labo PESCHANSKI, Créteil, France

Projet : Foetal neural transplant as a treatment for Huntington's disease : from pilot studies to clinical application (EuroHD-Phase II)



RETINA France, COLOMMIERS, France

Projet : Soutien aux *Projet*s de recherche sélectionnés par le Conseil Scientifique de Rétina France sur des thèmes de Thérapie génique et cellulaire.

ROTIG Agnès, Dr labo MUNNICH, Paris, France

Projet : Financement de 6 mois de CDD pour Sophie Lebon, technicienne chargée de la détection de mutations dans l'ADN mitochondrial, soit une somme de 15.000 euros

ROUSSET/ALLAMAND Jean-Pierre/Valérie, Dr labo JACQUET, Paris, France

Projet: Antibiotic-mediated readthrough of stop codon mutations responsible for muscular dystrophies pour financer une technicienne pour pallier l'absence de deux chercheuses en congé maternité

TABTI Nacira, Dr labo BARON-VAN EVERCOOREN, Paris, France

Projet: Pathophysiology of the Periodic Paralyses and Paramyotonia Congenita

TRANSGENE S.A., Strasbourg, France

Invitation des participants au protocole Transgène, patients et médecins, à une réunion le 18 juin 2003. (Prise en charge financière des patients).

VIOLLET/ZARHATE Louis/Mohamed, Dr labo MUNNICH, Paris, France

Projet: Prolongation du CDD d'un technicien pendant 1 an

VLM/VAINCRE LA MUCOVISCIDOSE, Paris, France

Projet : Financement des 16 Projets sélectionnés par le Conseil Scientifique de VLM dans la thématique « Thérapie génique et cellulaire »

VML/VAINCRE LES MALADIES LYSOSOMALES, Evry, France

Projet : Soutien à 6 subventions et 4 aides aux études sélectionnés par le Conseil Scientifique de VML dans la thématique « Thérapie génique et cellulaire »

Aide à l'organisation de colloques

BECHET Daniel, St Gènes Champanelle, France

Troisième colloque du Groupe Thématique "Protéolyse Cellulaire" de la Société Française de Biochimie et de Biologie Moléculaire., du 31 mars au 2 avril 2004 à Châtel-Guyon

BILLET Olivier, Montpellier, France

50 ans d'Adénovirus, en septembre 2003 à Montpellier

CHAPON Françoise, Caen, France



Les Premières Journées Annuelles de la Société Française de Myologie qui se tiendront à Caen les 9 et 10 Octobre 2003

CHELLY/AYADI Djamel/Hammadi, Paris, France

Organisation du 4ème cours théorique et pratique à la Faculté de Médecine de Sfax du 3 au 12 avril 2003.

COSSU Giulio, Milan, Italie

Gordon Research Conference on Myogenesis, organisée par G. Cossu à Il Ciocco, Italie, du 16 au 21 mai 2004.

DICKSON Georges, Londres, UK

11th Meeting of the European Society of Gene Therapy - Nov 14-17, 2003, Edinburg, Scotland

ENMC, Baarn, Hollande

Contribution à l'organisation des meetings organisés par l'ENMC

FAUGERON Godeleine, Paris, France

Organisation du Colloque 2003 de la Société Française de Génétique "Les Nouveaux Rôles de l'ARN" à l'Institut Pasteur 22-23 mai 2003.

FSH SOCIETY, Lexington, USA

Colloque organisé par S. Van der Maarel en novembre 2003 à Los Angeles

HILAIRE Gérard, Marseille, France

Colloque 2004 "Club du Motoneurone" organisé par G. HILAIRE à Marseille/Bastia du 26 au 28 février 2004

JORGENSEN Christian, Montpellier, France

3ème réunion annuelle de la Société Francophone de Thérapie Cellulaire et Génique les 19 et 20 juin 2003.

MILMAN Daniel, Paris, France

6ème colloque de Médecine et Psychanalyse : la fabrique des bébés ; passions pour l'embryon.

MONCKTON Darren, Glasgow, UK

4ème Conférence du Consortium International sur la Dystrophie Myotonique du 10 au 12 avril 2003 à Glasgow

PEARSON Christopher, Toronto, Canada

The 4th International Conference on Unstable Microsatellites and Human Disease in the winter of 2004



POUGET Jean, Marseille, France

XVIèmes Journées Neuromusculaires - les 11 et 12 septembre 2003 à Marseille.

PUCEAT/CHAUSSEPIED Michel/Patrick, Montpellier, France

32ème Congrès Européen du Muscle.

TEISSIE Justin, Toulouse, France

Université Européenne d'été 2003 : approches pluridisciplinaires de la Thérapie Génique à Toulouse du 1 au 5 septembre 2003

5/ APPEL D'OFFRES « MYOLOGIE FONDAMENTALE ET PATHOLOGIES DES MALADIES NEUROMUSCULAIRES »

Groupe A: Biologie intégrative du muscle

Aides aux études

DODILLE Magali, Dr labo ROSSI, Nice, France

Projet: Implication of chemotaxis in the survival and proliferation of myoblasts. Applications for optimizing cell therapies

JUNION Guillaume, Dr labo DASTUGUE, Clermont-Ferrand, France

Projet: Identification and analysis of ladybird target genes in Drosophila

KEDZIA Cécile, Dr labo KELLY, Paris, France

Projet: Effects of growth hormone on skeletal muscle - from in vivo to molecular studies

LAFUSTE/CHAZAUD Peggy, Dr labo GHERARDI, Créteil, France

Projet: Bone marrow-derived muscle satellite and mesenchymal cells. Evidence, kinetics and modulation in the normal and mdx mouse

MANFROID Isabelle, Dr labo PRADEL, Marseille, France

Projet : Functional analysis of the vertebrate teashirt gene family during somitogenesis and innervation of limb muscles



MORIN Christophe, Dr labo BARRITAULT, Créteil, France

Projet : Glycosaminoglycans in differentiating myoblasts in vitro and in regenerating muscle : effect of heparan sulfate mimetics (RGTA)

NGO-MULLER Valérie, Dr labo DAEGELEN, Paris, France

Projet: Mechanisms of skeletal muscle patterning during mouse limb development

NIEDERLANDER Nicolas, Dr labo DOREE, Montpellier, France

Projet : Développement d'un modèle cellulaire de cardiomyopathie dilatée ou hypertrophique

OUANOUNOU Gilles, Dr labo BAUX, Gif-sur-Yvette, France

Projet: Characterization and physiological role of the muscle inward rectifier K+ current during development of the embryonic neuromuscular junction.

PRODON/PATALANO François, Dr labo GACHE, Villefranche-sur-Mer, France

Projet: Role of cortical polarity and mRNA localization in primary muscle differenciation in ascidian

RENAULT Valérie, Dr labo BUTLER-BROWNE, Paris, France

Projet: Proliferative capacity of human skeletal muscle during aging : effects of exercice and oxidative stress on human satellite cells

SALLES Jérôme, Dr labo POUJEOL, Nice, France

Projet: Role of the MAP Kinase signalling pathways in muscle: involvement in Cachexia and degeneration

SARADAVEY Rathnam, Dr labo MANDEL, Illkirch, France

Projet: Comparative, molecular characterization of congenital and induced myopathies in the zebrafish embryo

SUELVES ESTEBAN Monica, Dr labo MUNOS CANOVES, Barcelone, Espagne

Projet : Mechanisms of action of the plasminogen activation system in skeletal muscle regeneration : role in mdx dystrophinopathy.

VU MANH Thien-phong, Dr labo PRADEL, Marseille, France

Projet : Functional genomics of Drosophila cardiogenesis : transcriptome analysis. Development of a relational database management system for data mining.

Subventions

BERNHEIM Laurent, Dr labo BERNHEIM, Genève, Suisse



Projet : Ionic channels, calcium signals, and differentiation factors during human myoblast fusion

BRESOLIN Nereo, Dr labo BRESOLIN, Milan, France

Projet : Muscle-derived stem cells from normal and dystrophic muscle tissues : a comparative study with clinical applications

BRUSTIS Jean-Jacques, Dr labo COTTIN, Talence, France

Projet: Calcium-dependent proteolysis during myoblast migration and fusion

BUCHERER GUILLERMIN Catherine, Dr labo LAURENT, Nouméa, Nouvelle Calédonie

Projet : Mechanical characterisation of human myoblasts. A new approach to study intermediate filament organisation

BUCKINGHAM Margaret, Dr labo BUCKINGHAM, Paris, France

Projet: Genes that regulate the initiation of myogenesis: Myf5 and Pax3 in the mouse embryo

BUCKINGHAM/MONTARRAS Margaret/Didier, Dr labo BUCKINGHAM, Paris, France

Projet: Role of Pax3 in relation to Pax7 in the specification of myosatellite cells and the search for adult muscle stem cells

BUTLER-BROWNE Gillian, Dr labo BUTLER-BROWNE, Paris, France

Projet: Human skeletal muscle: in health and disease

CABELLO Gérard, Dr labo CABELLO, Montpellier, France

Projet : Involvment of BTG1 in the regulation of myoblast differentiation and muscle development

COUX Olivier, Dr labo COUX, Montpellier, France

Projet: Roles of the ubiquitin-proteasome pathway in muscle differentiation

DANDOLO Luisa, Dr labo DAEGELEN, Paris, France

Projet: Study of Igf2 regulation by H19 using transgenic mouse models

DREYFUS Patrick A., Dr labo GHERARDI, Créteil, France

Projet : Bone marrow-derived muscle satellite and mesenchymal cells. Evidence, kinetics and modulation in the normal and mdx mice

DUPREZ Delphine, Dr labo MARO, Paris, France

Projet: Muscle and tendon interactions: Role of Fgf signalling and characterization of the Vertebrate homologues of Drosophila genes



FASANO Laurent, Dr labo PRADEL, Marseille, France

Projet : Functional analysis of the vertebrate teashirt gene family during somitogenesis and innervation of limb muscles

FERNANDEZ Anne, Dr labo MECHALI, Montpellier, France

Projet: Regulation of MyoD and role of Insulin-IGFs in myoblast differentiation and in activated satellite cells during regeneration

FONTAINE-PERUS Josiane, Dr labo FONTAINE PERUS, Nantes, France

Projet : Capacité des cellules embryonnaires à se différencier dans le muscle adulte

FONTAINE-PERUS Josiane, Dr labo FONTAINE-PERUS, Nantes, France

Projet: Myogenic potency of various embryonic cell categories explored in a mouse chick chimeric system

HAREL-BELLAN Annick, Dr labo HAREL-BELLAN, Villejuif, France

Projet: Involvement of chromatin modifiers in muscle specific promoter activation

JAGLA Krzysztof, Dr labo DASTUGUE, Clermont-Ferrand, France

Projet: Control of cell fate specification during muscle and heart formation

LEGER Jean, Dr labo ESCANDE, Nantes, France

Projet: Functional genomics of skeletal muscle in normal subjects and in patients presenting with neuromuscular disorders

LEIBOVITCH Serge Alexandre, Dr labo FEUNTEUN, Villejuif, France

Projet : biochemicals studies of the interaction between the myogenic factor MyoD and Atrogin-1, an E 3 ubiquitin ligase implicated in muscular atrophy

LI/DAEGELEN Zhenlin/Dominique, Dr labo PAULIN, Paris, France

Projet: Role of the serum response factor (SRF) in the cardiac, and skeletal muscles ontogenesis and function.

MAINA Flavio, Dr labo HENDERSON, Marseille, France

Projet: The HGF/MET signalling during muscle and motoneuron development (R dossier N; 8661 - AO 2002)

MAIRE Pascal, Dr labo DAEGELEN, Paris, France

Projet: Role of the six homeoproteins and their eya cofactors during muscle morphogenesis

MARCELLE Christophe, Dr labo PRADEL, Marseille, France

Projet: Mechanisms regulating myoblast differentiation in vertebrate embryos



MARIANI Jean, Dr labo MARIANI, Paris, France

Projet: Seeking the role of the orphan nuclear receptor RORalpha in muscle regeneration

MARTELLY Isabelle, Dr labo BARRITAULT, Créteil, France

Projet : Glycosominoglycans in differentiating myoblasts in vitro and in regenerating muscle : effect of heparan sulfate mimetics (RGTA).

MEGE René-Marc, Dr labo SOBEL, Paris, France

Projet : Cadherin-based cell adhesion in neuromuscular development. Molecular mechanims, signal transduction and cellular response

MUNOZ-CANOVES Pura, Dr labo MUNOZ-CANOVES, Barcelone, Espagne

Projet: Mechanisms of action of the plasminogen activation system in skeletal muscle regeneration: role in mdx dystrophinopathy.

PIETU Geneviève, Dr labo GIDROL, Evry, France

Projet : Transcriptional profiling of bone marrow and muscle-derived stem cells isolated by dye-efflux properties using DNA microarray

PUCEAT Michel, Dr labo DOREE, Montpellier, France

Projet : Ca2+ -dependent regulation of cardiac differentiation of normal and myopathic stem cell derived cardiomyocytes

ROBERT Benoît, Dr labo ROBERT, Paris, France

Projet: Role of Msx1 and Msx2 genes in muscle development and regeneration

ROSSI Bernard, Dr labo ROSSI, Nice, France

Projet : Implication of chemotaxis in the survival and proliferation of myoblasts. Applications for optimizing cell therapies. Demande faite sur 2 ans.

SARDET Christian, Dr labo GACHE, Villefranche-sur-Mer, France

Projet: Muscle differentiation in the ascidian embryo: roles of maternal determinants and asymmetric divisions. Demande faite sur 2 ans.

SEMERIVA Michel, Dr labo PRADEL, Marseille, France

Projet: Genetic control of cardiogenesis in Drosophila

SHI De-Li, Dr labo MARO, Paris, France

Projet : Analysis of regulatory cascade between Wnt signalling and myogenic factors in early myogenesis of vertebrate embryo

STRAEHLE Uwe, Dr labo MANDEL, Illkirch, France



Projet : Comparative, molecular characterization of congenital and induced myopathies in the zebrafish embryo. Demande faite sur 2 ans.

TAJBAKHSH Shahragim, Dr labo TAJBAKHSH, Paris, France

Projet: Characterisation of mouse muscle stem cells in the embryo and the adult

TORRENTE Yvan, Dr labo BRESOLIN, Milan, Italie

Projet : Identification of a putative pathway for the muscle homing of stem cells in a muscular dystrophy model

VINCENT Alain, Dr labo VINCENT, Toulouse, France

Projet : Muscle specification, fusion and differentiation in Drosophila; the collier-expressing muscle as a paradigm

Groupe B : Pathologies musculaires liées à des anomalies des protéines de structure

Aides aux études

ALVAREZ-FERNANDEZ/TEIJEIRA BAUTISTA Stela, Dr labo NAVARRO, Vigo, Espagne

Projet: Autosomal dominant congenital fiber type disproportion. linkage analysis in a large spanish family

BALGHI Haouaria, Dr labo RAYMOND, Poitiers, France

Projet : Calcium overload in dystrophin deficient muscle cells : involvement of Ca++ releases from the sarcoplasmic reticulum ?

BENAYOUN Béatrice, Dr labo DANOS, Evry, France

Projet: Expression profiling in calpainopathies

BOITTIN François-Xavier, Dr labo RUEGG, Lausanne, Suisse

Projet: Nerve-muscle co-culture to study calcium regulation in a murine model of DMD

BUJ-BELLO Anna, Dr labo MANDEL, Illkirch, France

Projet: Molecular physiopathology and gene transfer in X-linked myotubular myopathy

DIOGON Marie, Dr labo MANDEL, Illkirch, France

Projet : Use of C. elegans as a model to study the mechanisms leading to Epidermolysis Bullosa Simplex with Muscular Dystrophy (MD-EBS)



DISSET Antoine, Dr labo CLAUSTRES, Montpellier, France

Projet: Analysis of mechanisms underlying normal and abnormal regulation of dystrophin pre-mRNA splicing

DONNER Kat, Dr labo DE LA CHAPELLE, Helsinki, Finlande

Projet: The role of nebulin and beta-tropomyosin mutations in nemaline myopathy

FRAYSSE Bodvaë, Dr labo COMTE-CAMERINO, Bari, Italie

Projet : Evaluation of different drug treatments on skeletal muscle Ca2+ homeostasis of exercised MDX mice

LANTERI-MINET (**LIPSKAIA**) **Larissa**, Dr labo FISCHMEISTER, Chatenay-Malabry, France

Projet: Role of the sarcoplasmic reticulum in the mechanism of development of vascular dysfunction in cardiomyopathy

LAPOUGE Xarine, Dr labo BORRADORI, Genève 14, Suisse

Projet : Characterization of the binding partners of human synemin, a novel intermediate filament protein that associate with desmin in striated muscle cells

MANSOUR Sana, Dr labo PORTERO, Paris, France

Projet: approche dissociée de la mobilité des membres inférieurs. Application à l'analyse des déficits et des asymétries fonctionnelles.

MAUGENRE Svetlana, Dr labo GUICHENEY, Paris, France

Projet : Identification of new genes responsible for merosin-positive congenital muscular dystrophies

RAYNAUD Fabric, Dr labo VIAL, Montpellier, France

 $Projet: \mu$ -calpain translocation to gamma filamin: identification of the binding interface and development of vectorized peptides to inhibit the process.

TRUCHET Sandrine, Dr labo HENRY, Paris, France

Projet : Plasma membrane repair mechanism in myocytes and pathophysiology of muscular dystrophies

Subventions

ALLARD Bruno, Dr labo ALLARD, Villeurbanne, France

Projet : Characterization of Ca 2+ entry pathways in skeletal muscle fibers from normal and mdx mice



BENYAMIN Yves, Dr labo VIAL, Montpellier, France

Projet : μ -calpain translocation to muscle gamma filamin : interface indentification and development of molecular tools to explore filamin- and calpain- related LGMDs. Calpain distribution and participation beside ...

BERDEAUX Alain, Dr labo BERDEAUX, Le Kremlin-Bicêtre, France

Projet: Volume-dependent chloride channels and apoptosis in cardiomyocytes issued from mdx mice and GRMD canine myocardium

BLOT Stéphane, Dr labo BLOT, Maisons-Alfort, France

Projet : Development and genetic analysis of a new model of dystrophinopathy in a Labrador retriever strain

BONNE Gisèle, Dr labo GUICHENEY, Paris, France

Projet: Genetics and physiopathology of laminopathies affecting the striated muscles

BRAIS Bernard, Dr labo BRAIS, Montréal, Quebec, Canada

Projet : Clinical, pathological and molecular characterization of severe OPMD and OPMD-like phenotypes

BUSHBY Kate, Dr labo STRACHAN, Newcastle upon Tyne, UK

Projet: Proteomic Approaches to Understanding Dysferlinopathy

CARRIER Lucie, Dr labo GUICHENEY, Paris, France

Projet : In vivo and ex vivo functional analysis of cardiac myosin binding-protein C mutants implicated in familial hypertrophic cardiomyopathy

CARTAUD Jean, Dr labo KARSENTI, Paris, France

Projet: Subsynaptic differentiation at the Vertebrate Neuromuscular Junction

COGNARD Christian, Dr labo RAYMOND, Poitiers, France

Projet : Role of organelles-related mechanisms in disturbance of calcium handling in dystrophin deficient muscle cells

DARCHEN François, Dr labo HENRY, Paris, France

Projet : Plasma membrane repair mechanism in myocytes and pathophysiology of muscular dystrophies

GOUBEL Francis, Dr labo BARTHES-BIESEL, Compiègne, France

Projet : Modelling of the in vivo human muscle force-velocity relationship: Application to DMD children

GUICHENEY/KROL Pascale/Alain, Dr labo GUICHENEY, Paris, France



Projet : Genetic congenital muscular dystrophies : localisation and function of the selenoprotein N

JASMIN Bernard, Dr labo JASMIN, Ottawa, Ontario, Canada

Projet : Molecular Regulation of Utrophin in Skeletal Muscle fibers : Role of alternate 5'UTRs, nitric oxide signaling and DNA methylation

LABOUESSE Michel, Dr labo MANDEL, Illkirch, France

Projet : C. elegans as a model to investigate human pathologies associated with the loss of plakins

LACOLLEY Patrick, Dr labo LAURENT, Paris, France

Projet : Cardiovascular phenotype in mice lacking alpha 1 beta 1 integrin and signalization in vascular smooth muscle

LE RUMEUR Elisabeth, Dr labo LE RUMEUR, Rennes, France

Projet: Interaction of dystrophin rod domain with membrane phospholipids. Analysis and molecular characterization with the 2nd repeat and a mini rod domain

LOMPRE Anne-Marie, Dr labo FISCHMEISTER, Chatenay-Malabry, France

Projet: Role of the sarcoplasmic reticulum in the mechanism of development of vascular dysfunction in cardiomyopathy

MANDEL/LAPORTE Jean-Louis/Jocelyn, Dr labo MANDEL, Illkirch, France

Projet: Function and muscular specificity of the protein implicated in myotubular myopathy and gene transfer assays in mouse

MARALDI Nadir Mario, Dr labo MARALDI, Bologne, Italie

Projet : Electron microscopy and immunocytochemical study of collagen VI and collagen VI-related proteins in UCMD

MEUNE/DUBOC Christophe/Denis, Dr labo WEBER, Paris, France

Projet: Tissue doppler echocardiography in dystrophinopathy and detection of early cardiac dysfunction in female carriers

MIRONNEAU Jean, Dr labo MIRONNEAU, Bordeaux, France

Projet : Role of caveolin-3 and sarcoglycans in Ca 2+ signalling of smooth muscle myocytes from genetically modified mice

NAVARRO Carmen, Dr labo NAVARRO, Vigo, Espagne

Projet : Autosomal dominant congenital fiber type disproportion. Linkage analysis in a large Spanish family

PANTHIER Jean-Jacques, Dr labo PANTHIER, Maisons-Alfort, France



Projet : Genetic and patholophysiological study of a hereditary centronuclear myopathy in the dog

PAULIN Denise, Dr labo PAULIN, Paris, France

Projet : Myofibrillar myopathies : function of synemin isoforms in desmin-related myopathies

PEROT Chantal, Dr labo BARTHES-BIESEL, Compiègne, France

Projet: Structural and functional assessment of a muscle spindle plasticity

PICAUD/RENDON Serge/Alvaro, Dr labo SAHEL, Paris, France

Projet : Functional role of dystrophins in synaptic transmission by patch-clamp recording of retinal neurons in mdx3cv mice

RUEGG Urs, Dr labo RUEGG, Lausanne, Suisse

Projet: Investigation of the cytoprotective mechanisms of creatine and of green tea extract in dystrophic muscle

SAKANYAN Vehary, Dr labo SAKANYAN, Nantes, France

Projet : Study of a network of molecular interactions implicated in neuromuscular pathologies by protein array.

SEGALAT Laurent, Dr labo COUBLE, Villeurbanne, France

Projet: Genetic analysis of genes interacting with dystrophin: studies in C. elegans and mouse models

TOMA Flavio, Dr labo TOMA, Evry, France

Projet : NMR Investigations of the structure and Functional ZZ domain of human dystrophin : role of the pathogenic mutations

TOPALOGLU Haluk, Dr labo KALE, Ankara, Turquie

Projet: Study of neuromuscular disorders in Turkey and establishing a nation-wide database network, part II

TUFFERY-GIRAUD Sylvie, Dr labo CLAUSTRES, Montpellier, France

Projet : Analysis of mechanisms underlying normal and abnormal regulation of dystrophin pre-mRNA splicing

UDD Bjarne, Dr labo GROOP, Helsinki, Finlande

Projet: Tibial muscular dystrophy (TMD)/LGMD2J - gene defects and functional genomics

VAILLEND Cyrille, Dr labo LAROCHE, Orsay, France



Projet : Cellular and molecular mechanisms of the cognitive impairments in dystrophinopathies

VICART Patrick, Dr labo BUTLER-BROWNE, Paris, France

Projet: Molecular and cellular function of desmin-associated proteins in normal and pathologic muscular cells

VICART Patrick, Dr labo BUTLER-BROWNE, Paris, France

Projet: Desmin and alpha B-crystallin gene analysis in Desmin-Related Myopathies

Groupe C: Motoneurone et jonction neuromusculaire

Aides aux études

AGHAIE Asadollah, Dr labo GUENET, Paris, France

Projet: Positional cloning of an original mouse mutation with neuropathological effects

AZZEDINE Hamid, Dr labo HIRSCH, Paris, France

Projet : Etude génétique des formes autosomiques recessives de la maladie de Charcot-Marie-Tooth (ARCMT)

CARDENAS MATUS Julio Cesar, Dr labo BAUX, Gif-sur-Yvette, France

Projet : IP3 - receptors and associated Ca2+ - signals at the neuromuscular junction and skeletal muscle components

CATONE Christelle, Dr labo TERNAUX, Marseille, France

Projet: Morphogenic and trophic effects of acetylcholine on embryonic cultured rat spinal motoneurons

DE SANDRE-GIOVANNOLI Annachiara, Dr labo LEVY, Marseille, France

Projet : Physiopathology of autosomal recessive HMSN due to mutations in human and mouse LMNA, and human GDAP1 and MTMR2

DOUIN-ECHINARD Victorine, Dr labo GUERY, Toulouse, France

Projet : Estrogens enhance TH1 responses in vivo. Implication in the pathogenesis of auto-immune myasthenia gravis

FARAUT Brice, Dr labo GUICHENEY, Paris, France

Projet: Role of thrombin, its receptor PAR-1and its inhibitor PN-1 in the formation and plasticity of the neuromuscular junction



GEIB Sandrine, Dr labo HENDERSON, Marseille, France

Projet : Role of axonal transport of SMN, and mRNA in motoneurons : effects of deletion of exon 7

GIRARD Cyrille, Dr labo JEANTEUR, Montpellier, France

Projet : Dissection of SMN and hTGS1 functions in RNPs assembly and Cajal body formation using RNA interference

MARTHIENS Véronique, Dr labo SOBEL, Paris, France

Projet : Roles of type II cadherins 6 and 11 in spinal motoneurons differentiation and survival

MERAOUNA Amel, Dr labo BERRIH-AKNIN, Le Plessis Robinson, France

Projet : Mechanism of action therapeutical agents : functional and transcriptome analysis

MOUROT Alexandre, Dr labo GOELDNER, Illkirch, France

Projet : Structure and dynamic of muscular acetylcholine receptor as studied by photoaffinity labeling

NANCY Patrice, Dr labo RENAUD, Le Plessis Robinson, France

Projet : Role of sexual hormones in the development of thymic hyperplasia in myasthenia gravis

RENAULT Ludovic, Dr labo ROCHAT, Marseille, France

Projet : Allosteric regulation of acetylcholinesterase catalysis by monoclonal antibodies

ROUX Sylvie, Dr labo BRULET, Paris, France

Projet: Characterization of the neurotrophic signal in the nerve-muscle system by use of the C-Francegment of the tetanus toxin

SOARES Sylvia, Dr labo CALAS, Paris, France

Projet : Functional analysis of the microtubule-associated protein 1B in peripheral regeneration and myelination

STROCHLIC Laure, Dr labo CARTAUD, Paris, France

Projet: Role of the MuSK complex in the signaling at the vertebrate neuromuscular junction

TALY Antoine, Dr labo CHANGEUX, Paris, France

Projet : Structural investigations of the conformational transitions of nicotinic receptors



VANDIEDONCK Claire, Dr labo BACH, Paris, France

Projet : Fine mapping and identification of the HLA-linked MYAS1 locus in myasthenia gravis patients with thymus hyperplasia

YU Hua, Dr labo BAUX, Gif-sur-Yvette, France

Projet: Biophysical and pharmacological analysis of the mutant AChR with congenital myasthenic syndrom (CMS)

Subventions

AVNER Philip, Dr labo AVNER, Paris, France

Projet : Control of cell proliferation by the "nucleosome assembly protein 1 like 2 " : a tool for neuronal regeneration ?

BELIN/ANTOINE Marie-Françoise/Jean-Christophe, Dr labo BELIN, Lyon, France

Projet : Peripheral nerve survival and regenerescence : role of collapsin response mediator proteins

BERRIH-AKNIN Sonia, Dr labo RENAUD, Le Plessis Robinson, France

Projet: Myasthenia gravis network

BERRIH-AKNIN Sonia, Dr labo RENAUD, Le Plessis Robinson, France

Projet: Pathophysiological mechanisms of myasthenia gravis

BESSEREAU Jean-Louis, Dr labo TRILLER, Paris, France

Projet : Genetic analysis of acetylcholine receptor metabolism in the model system Caenorhabditis elegans

BORDONNE Rémy, Dr labo JEANTEUR, Montpellier, France

DOSSIER COMPLET: Dissection of SMN and hTgs1 functions in RNPs assembly and Cajal body formation

BRENNER Talma, Dr labo BRENNER, Jérusalem, Israël

Projet : Inhibition of acetylcholinesterase by antisens oligonucleotides in myasthenia gravis and experimental autoimmune myasthenia gravis

BRICE Alexis, Dr labo HIRSCH, Paris, France

Projet : Clinico-genetic analysis of complex forms of hereditary neuropathies with pyramidal signs and/or cerebellar ataxia

BRULET Philippe, Dr labo BRULET, Paris, France

Projet : Analysis of membrane traffic at the mouse neuromuscular junction



CATALA Martin, Dr labo BUTLER-BROWNE, Paris, France

Projet : The molecular pathway involved in blocking differentiation of motoneuronal stem cells in the spinal cord

CHARBONNIER Frédéric, Dr labo VIDAL, Paris, France

Projet: Effects of Physical exercise on the motor behaviour of amyotrophic-lateral-sclerosis mice

CHARNAY Patrick, Dr labo CHARNAY, Paris, France

Projet : Molecular genetics of myelinisation and of peripheral nervous system development

COHEN-KAMINSKY Sylvia, Dr labo BERRIH-AKNIN, Le Plessis Robinson, France

Projet : Regulatory antibodies to the T cell receptor in myasthenia gravis : towards a potential role in T cell homeostasis

COURAUD François, Dr labo SEAGAR, Marseille, France

Projet : Developmental expression of the Nav1.6 sodium channel in the normal and pathologic neuromuscular junction in mice

DELAPEYRIERE Odile, Dr labo HENDERSON, Marseille, France

Projet : Characterization of subpopulations of motoneuron affected in neurodegenerative diseases

DUBOURG Odile, Dr labo HIRSCH, Paris, France

Projet: Molecular genetics of spinal Charcot-Marie_Tooth disease

EYER Joël, Dr labo EYER, Angers, France

Projet : Analysis of neurofilaments/tubulin interactions and their pathological perturbations

FALEMPIN Maurice, Dr labo FALEMPIN, Villeneuve d'Ascq, France

Projet: Deafferentation and muscular consequences

FUCHS/ SOUROUJON Sara/Miriam, Dr labo FUCHS, Rehovot, Israel

Projet: Immunoregulation of Experimental Autoimmune Myasthenia Gravis

GALLI Thierry, Dr labo GIRAULT, Paris, France

Projet: Role of membrane traffic in neuronal differentiation and plasticity

GASCAN Hughes, Dr labo GASCAN, Angers, France

Projet : Molecular and functional characterization of CNTF-3, a novel neurotrophic cytokine



GIRAULT Jean-Antoine, Dr labo GIRAULT, Paris, France

Projet: Molecular basis of axo-glial contacts in peripheral nerves

GUENET Jean-Louis, Dr labo GUENET, Paris, France

Projet : Positional cloning of an original mouse mutation with neuropathological effects

GUERY Jean-Charles, Dr labo DRUET, Toulouse, France

Projet : Estradiol promotes the development of antigen-specific Th1 cells in vivo. Implication in the pathogenesis of Myasthenia gravis

HAASE Georg, Dr labo ROUGON, Marseille, France

Projet: Molecular and cellular mechanisms of axonal degeneration

HANTAI/EYMARD Daniel/Bruno, Dr labo GUICHENEY, Paris, France

Projet: Pathophysiological and molecular characterization of congenital myasthenic syndromes

HENDERSON Christopher, Dr labo HENDERSON, Marseille, France

Projet : Factors regulating survival and growth of neurons in spinal cord and peripheral nerve

HIBERT Florence, Dr labo GOELDNER, Illkirch, France

Projet : Original chemical tools for structural and dynamic investigations on the nicotinic acetylcholine receptor

KATO Ann C., Dr labo KATO, Genève 4, Suisse

Projet : Studies of motoneuron degeneration in animal models : cell replacement therapy and gene profiling of degenerating motoneurons

KREJCI Eric, Dr labo ASCHER, Paris, France

Projet : Synapse organization and cholinergic transmission at the neuromuscular junction

LE GUERN Eric, Dr labo HIRSCH, Paris, France

Projet : Expression of the proteins encoded by the genes involved in autosomal recessive forms of dysmyelinating CHARCOT MARIE TOOTH disease during development and myelination in Human

LEFEBVRE Suzie, Dr labo KARSENTI, Paris, France

Projet : Biochemical and cellular characterization of the Spinal Muscular Atrophy gene product, SMN



LEVY Nicolas, Dr labo FONTES, Marseille, France

Projet: Physiopathology of autosomal recessive HMSN due to mutations in human and mouse LMNA, and human GDAP1 and MTMR2.

MANDEL/PUJOL Jean-Louis/Aurora, Dr labo MANDEL, Illkirch, France

Projet : Characterization, functional analysis and therapy for mouse models of X-linked adrenomyeloneuropathy

MARCHOT Pascale, Dr labo ROCHAT, Marseille, France

Projet : Allosteric regulation of acetylcholinesterase catalysis by monoclonal antibodies

MASSOULIE Jean, Dr labo ASCHE, Paris, France

Projet: Molecular and celllar biology of cholinesterases

MOLGO Jordi, Dr labo BAUX, Gif-sur-Yvette, France

Projet : IP3 - receptors and associated Ca2+ -signals at the neuromuscular junction and skeletal muscle components

PETTMANN/HAASE Brigitte/Georg, Dr labo HENDERSON, Marseille, France

Projet: Motor-neuron specific cell death pathways triggered by ALS-linked SOD1 mutations

ROUGON Geneviève, Dr labo ROUGON, Marseille, France

Projet: Peptides and peptido-mimetics for cell adhesion molecules: evaluation of their therapeutic potential for spinal cord repair.

SCHAEFFER Laurent, Dr labo JALINOT, Lyon, France

Projet: Chromatin modifications and neuromuscular differentiation

SEAGAR Michael, Dr labo SEAGAR, Marseille, France

Projet : Regulation of neurotransmitter release by the synaptobrevin/ synaptophysin complex

TERNAUX Jean-Pierre, Dr labo TERNAUX, Marseille, France

Projet: Influences of acetylcholine, serotonin and noradrenalin on the development of motoneurones and neuromuscular junctions

TRILLER Antoine, Dr labo TRILLER, Paris, France

Projet: Molecular genetic of synaptogenesis: afferent innervation of the motoneuron



VANDROMME Marie, Dr labo ALMARIC, Toulouse, France

Projet: Presenilin dependent intramembrane cleavage of erbB4 at the neuromuscular junction

VIGNY Marc, Dr labo SOBEL, Paris, France

Projet : ALK receptor in the neuromuscular system : expression and mechanisms of action

VIOLLET Louis, Dr labo MUNNICH, Paris, France

Projet : Genetic mapping and gene identification for autosomal recessive chronic distal spinal muscular atrophies

Groupe D: Canalopathies, Mitochondriopathies, Maladies Inflammatoires et autres Pathologies neuromusculaires

Aides aux études

BELANGER Guy, Dr labo JASMIN, Ottawa, Ontario, Canada

Projet: Role of the RNA-Binding protein Staufen at the neuromuscular junction: possible link with myotonic dystrophy

BIDAUD Isabelle, Dr labo DEMAILLE, Montpellier, France

Projet: The role of T-type calcium channels in muscle differentiation

CARVAJAL AGUILERA Karla, Dr labo MORENO-SANCHEZ TLALPAN, Mexico D.F., Mexico

Projet: Alterations in cardiac energy metabolism in the AMPK deficient mouse

DUVAL Carine, Dr labo VILLARROYA, Barcelona, Spain

Projet: Impact of UCP-3 on mithochondria: bioenergetic metabolism and reactive oxygen species. Implication in myopathy

FERNANDES Fanny, Dr labo SEAGAR, Marseille, France

Projet : Physiopathology of neuromuscular communication : molecular dissection of the voltage-dependant sodium channels

LAGIER-TOURENNE Clotilde, Dr labo MANDEL ILLKIRCH CEDEX France

Projet : Physiopathological implications of thymidine levels in Mitochondrial NeuroGastroIntestinal Encephalomyopathy

LHUILLIER Loïc, Dr labo BARON VAN EVERCOOREN, Paris, France



Projet : Molecular and cellular pathophysiology of hypokalemic periodic paralysis type 2

MAGUY Ange, Dr labo MICHEL, Paris, France

Projet: MAGUKs proteins and plasticity of neronal synapse and cardiac gap junctions

MALLOUK Nora, Dr labo LAZDUNSKI, Valbonne, France

Projet : The 2P domain K+ channel TREK-1 in motoneurons : regulation by metabotropic glutamate receptors and the role of protein kinase C

MENTEYNE Alexis, Dr labo BAUX, Gif-sur-Yvette, France

Projet : Studies of the novel Calcium-releasing messengers NAADP in motoneurones : measurement of endogeneous NAADP levels

ROLLAND Jean-François, Dr labo CONTE CAMERINO, Bari, Italie

Projet: Imaging and patch-clamp evaluation of drug treatments on altered Ca 2+ hemeostasis of exercised MDX mouse muscles

SCHWIMMER Christine, Dr labo DI RAGO, Bordeaux, France

Projet : Yeast as a model for research on mutations in the mitochondrial ATP6 gene associated with human pathologies

SHESTAKOVA Elena, Dr labo DJIAN, Paris, France

Projet: Role of nuclear spatial organization and chromatin structure during the establishment of myotonic dystrophy

Subventions

BARHANIN Jacques, Dr labo LAZDUNSKI, Valbonne, Sophia-Antipolis, France

Projet : Skeletal muscle K+ channels: functional properties and consequences of periodic paralysis mutations

BELAYEW Alexandra, Dr labo BELAYEW, Mons, Belgique

Projet: Study of the DUX4 protein encoded by a candidate gene for FSHD

CANCELA José-Manuel, Dr labo BAUX, Gif-sur-Yvette, France

Projet : Studies of the novel Ca 2+ releasing messengers NAADP : characterization of its physiological roles in motoneurone and other cell types

CASAS François, Dr labo CABELLO, Montpellier, France

Projet: Involvment of the direct T3 mitochondrial pathway in mouse muscle development



CHARNET Pierre, Dr labo DOREE, Montpellier, France

Projet: Calcium channels and nicotinic receptors in normal and pathologic synaptic transmission

CORRAL-DEBRINSKI Marisol, Dr labo DREYFUS, Paris, France

Projet: The cellular localization of mRNAs encoding mitochondrial proteins when altered could be involved in a metabolic myopathies

DARGENT Bénédicte, Dr labo SEAGAR, Marseille, France

Projet: Physiopathology of neuromuscular communication: molecular dissection of the clustering motif of the voltage-dependant sodium channel

DERIJARD Benoît, Dr labo POUJEOL, Nice, France

Projet: Cell signalling in myotonic dystrophy skeletal muscle

DI RAGO Jean-Paul, Dr labo AIGLE, Bordeaux, France

Projet: Genetic analysis of yeast ATP synthase and human pathologies

DUJARDIN/BONNEFOY Geneviève/Nathalie, Dr labo AGGERBECK, Gif-sur-Yvette, France

Projet: Nuclear control of mitochondrial functions from yeasts to man

FONTAINE Bertrand, Dr labo BARON, Paris, France

Projet : Study of the relationship between basal lamina defect and muscle hyperexcitability : construction of a mouse model for Schwartz-Jampel symdrome

FURLING/BUTLER-BROWNE Denis/Gillian, Dr labo BUTLER-BROWNE, Paris, France

Projet : Determination of molecular alterations involved in the muscle physiopathology of DM1 using a human in vitro model

GOURDON Geneviève, Dr labo JUNIEN, Paris, France

Projet : Transgenic mice models for myotonic dystrophy : understanding the physiopathology towards therapeutic tools

GOURDON Geneviève, Dr labo JUNIEN, Paris, France

Projet : CTG repeat instability in myotonic dystrophy: understanding the mechanism(s) of expansion as a route towards therapy

HAMSHERE/ARMOUR Marion/John, Dr labo HAMSHERE, Nottingham, UK

Projet : Myotonic dystrophy: could a change in direction of DNA replication make the repeat unstable?



HATEM Stéphane, Dr labo MICHEL, Paris, France

Projet: Role of the anchoring proteins MAGUK in the organization and properties of neuronal and cardiac channels

LACAMPAGNE Alain, Dr labo VASSORT, Montpellier, France

Projet: Effects of statins on Ca2+ homeostasis in human skeletal muscle

LAZDUNSKI Michel, Dr labo LAZDUNSKI, Valbonne, France

Projet: Asic channels: Role in pain perception and in brain pathologies

LEVY Nicolas, Dr labo FONTES, Marseille, France

Projet : Characterization of the genetics basis for congenital fibrosis of extra ocular muscles type 4 (CFEOM4), a non progressive ocular neuromuscular disorder

LOMBES/ROJO Anne/Manuel, Dr labo GUICHENEY, Paris, France

Projet: Molecular investigations of human mitochondrial myopathies

LUNARDI/MARTY Joël/Isabelle, Dr labo LUNARDI, Grenoble, France

Projet : Study of the skeletal muscle calcium release complex in normal situation and in congenital myopathies associated with cores.

MITRANI-ROSENBAUM Stella, Dr labo GALUN, Jérusalem, Israël

Projet: The pathological mechanism of hereditary inclusion body myopathy-proteomic approach

MONCKTON/JOHNSON Darren G./Keith J., Dr labo JOHNSON, Glasgow, UK

Projet: Determining the function of SIX5 and its contribution to the phenotype of myotonic dystrophy (DM1)

MORRIS Glenn E., Dr labo MORRIS, Wrexham, UK

Projet: Monoclonal antibodies to human "muscleblind" proteins for studies of myotonic dystrophy and human development

MORRIS Glenn E., Dr labo MORRIS, Wrexham, UK

Projet : Monoclonal antobodies to human CUG-binding proteins for studies of myotonic dystrophy

NARGEOT Joël, Dr labo BOCKAERT, Montpellier, France

Projet: Calcium channels and muscular physiology and pathophysiology

OZDEN Simona, Dr labo GESSAIN, Paris, France

Projet: Pathogenesis of muscle injury in myositis associated with the human retrovirus HTLV-1



PLONQUET Anne, Dr labo GHERARDI, Creteil, France

Projet : Evaluation of cellular cytotoxic immunity in polymyositis associated with anti-JO-1 antibodies

PUYMIRAT Jack, Dr labo PUYMIRAT, Quebec, Canada

Projet : Identification of a factor produced by myotonic dystrophy myoblasts that inhibits myogenic differentiation

ROTIG Agnès, Dr labo MUNNICH, Paris, France

Projet: Mitochondrial respiratory chain diseases of nuclear origin

SERGEANT Nicolas, Dr labo BEAUVILLAIN, Lille, France

Projet: Identification of nuclear factors responsible for the trans-effect of myotonic dystrophy of type I mutation

TUPLER Rossella Ginevra, Dr labo SCAPPATICCI, Pavia, Italie

Projet: The molecular basis of facioscapulohumeral muscular dystrophy

VENTURA-CLAPIER Renée, Dr labo FISCHMEISTER, Chatenay Malabry, France

Projet : Functional consequences on cardiac and skeletal muscle metabolism of AMP kinase deficiency

ZINN-JUSTIN Sophie, Dr labo MENEZ, Gif-sur-Yvette, France

Projet: Structural characterization of emerin and lamin A/C, two nuclear envelope proteins mutated in muscular dystrophies

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