SF-ENMG
Société Francophone d'ElectroNeuroMyoGraphie

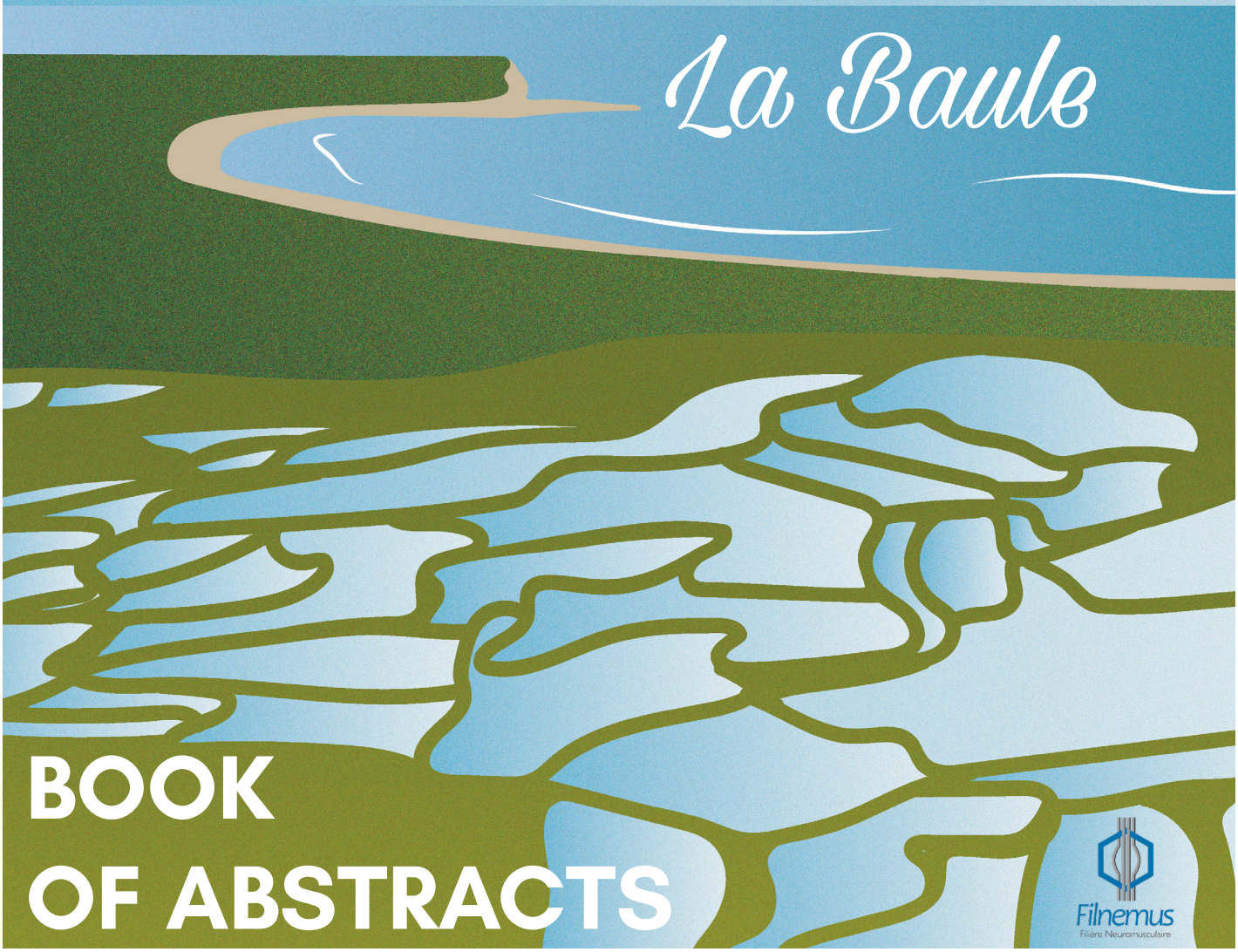
13th International Congress
of Paediatric Electromyography

PAEDIATRIC EMG

November 13-15, 2023

Palais des Congrès - Atlantia, La Baule

ORGANIZING COMMITTEE
Cyril Gitiaux, Armelle Magot, Yann Péréon



BOOK OF ABSTRACTS

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MONDAY, NOVEMBER 13TH

08:15 - 08:45 am **Participants Welcoming**

SESSION 1 - BASICS WITH CHILDREN

Chairmen: Matthew Pitt, Yann Péréon

08:45 - 09:00 am Introduction
Yann Péréon - Nantes, France

09:00 - 09:30 am Engaging the children: How I perform EDX exam in children
Matthew Pitt - London, UK

09:30 - 10:00 am Nerve conduction velocities & motor unit potentials: What is different in children?
Joe F. Jabre - Los Angeles, USA

10:00 - 10:30 am How I analyse MUP when doing needle EMG
Sanjeev D. Nandedkar - Middleton, USA

10:30 - 11:00 am **Coffee break with partners**

SESSION 2 - CLASSICAL SITUATIONS

Chairmen: Nancy Kuntz, Cyril Gitiaux

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Jacquie Deeb - London, UK

11:30 - 12:00 pm How I perform EDX exam when facing brachial plexus obstetrical palsy
Ruth Van der Looven - Gent, Belgium

12:00 - 12:30 pm How I perform EDX exam when facing scapula alata
Yann Péréon - Nantes, France

12:30 - 01:30 pm **Symposium**
Chairman: Yann Péréon
LUPIN NEUROSCIENCES
Can EMG be supportive in timely diagnosis of myotonic disorders?
Yann Péréon, Emma Matthews, Valeria Sansone

12:30 - 02:15 pm **Lunch Break with partners**

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SCIENTIFIC PROGRAM

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SESSION 3 - NEUROMUSCULAR JUNCTION

Chairmen: Peter Kang, Véronique Manel

02:15 – 02:45 pm How I perform EDX exam facing possible neuromuscular junction disorder
Matthew Pitt – London, UK

02:45 – 03:15 pm Congenital myasthenic syndromes: New genes, new EDX patterns
Tanya Stojkovic – Paris, France

03:15 – 03:45 pm New insights in Myasthenia Gravis treatment
Emmanuelle Campana-Salort – Marseille, France

03:45 - 04:15 pm Coffee break with partners & Posters

SELECTED ORAL COMMUNICATIONS

Chairwomen: Jacquie Deeb, Armelle Magot

04:15 – 04:30 pm *Muscle ultrasound as a complementary diagnostic value in neuromuscular diseases*
Justyna Pigońska – Lodz, Poland & Marta Banach, Cracow, Poland

04:30 – 04:45 pm *Correlation of nerve ultrasound findings with clinical and electrophysiological characteristics in Paediatric Charcot-Marie-Tooth patients*
Hanna Küpper – Tübingen, Germany

04:45 – 05:00 pm *Median nerve ultrasound as a screening tool diagnosing carpal tunnel syndrome in mucopolysaccharidosis type 1H patients*
Peter Karachunski – Minneapolis, USA

05:00 – 05:15 pm *Carpal tunnel syndrome in pediatric patients with PIK3 pathway abnormalities*
Arnaud Chefdor – Angers, France

05:15 – 05:30 pm *ENMG in paediatric ICU*
Laura Nastasi – London, UK

05:30 – 05:45 pm *Polyneuropathy in vincristine treated pediatric ALL patients*
Leena Lauronen – Helsinki, Finland

05:45 – 06:00 pm *Electrophysiological characterization of essential tremor in children and adolescents*
Julie Piarroux – Montpellier, France

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SCIENTIFIC PROGRAM

TUESDAY, NOVEMBER 14TH

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SESSION 4 - NEUROPATHIES 1

Chairmen: Hugh McMillan, Susana Quijano Roy

09:00 - 09:30 am How I perfom EDX exam when facing ataxic gait or balance disorder
Cyril Gitiaux - Paris, France

09:30 - 10:00 am How I perform EDX exam when facing gait disorder with foot deformity
John McHugh - Dublin, Eire

10:00 - 10:30 am CMT genetics: Clinical & electrophysiological correlations
Jean-Baptiste Noury - Brest, France

10:30 - 11:00 am **Coffee break with partners & Posters**

SESSION 5 - MYOPATHIES

Chairmen: John McHugh, Emmanuelle Campana Salort

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Peter Kang - Minneapolis, USA

11:30 - 12:00 pm How I perform EDX exam when facing exercise intolerance
Véronique Manel - Lyon, France

12:00 - 12:30 pm EDX vs muscle imagery (MRI, US) in children: Pros & Cons
Susana Quijano Roy
& Amal Oulhissane-Omar - Garches, France

12:30 - 01:30 pm **Symposium**
Chairman: Yann Péréon
NOVARTIS
SMA: How can the ENMG help in the therapeutic decision?
Juliette Ropars, Jean-Baptiste Davion, Rémi Barrois

12:30 - 02:15 pm **Lunch Break with partners**

TUESDAY, NOVEMBER 14TH

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Chairmen: Joe Jabre, Tanya Stojkovic

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Hugh McMillan - Ottawa, Canada

02:45 - 03:15 pm New antibodies in dysimmune neuropathies
Jérôme Devaux - Montpellier, France

03:15 - 03:45 pm Small fibres in 2023:
How to explore, pathophysiology, new diseases or concepts?
Nancy Kuntz - Chicago, USA

03:45 - 04:15 pm EDX in veterinarian practice
Jean-Laurent Thibaud - Créteil, France

04:15 - 04:45 pm Coffee break with partners & Posters

WORKSHOPS - FIRST SESSION

Room	Pierre Sastre	Oreaux Baguenaud	Euler-Lancastria
04:45 - 05:30pm	WS1 Single fibre EMG *Matthew Pitt*	WS2 Diaphragm *Yann Péréon*	WS3 Munix *Sanjeev D. Nandedkar*
05:45 - 06:30pm	WS1 Single fibre EMG *Matthew Pitt*	WS2 Diaphragm *Yann Péréon*	WS3 Munix *Sanjeev D. Nandedkar*

07:00 pm CONGRESS DINNER

PAEDIATRIC EMG

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WEDNESDAY, NOVEMBER 15TH

08:30 - 09:00 am Participants Welcoming

SESSION 7 - NEUROPATHIES 3

Chairmen: Ruth Van der Loeven, Jean-Baptiste Noury

09:00 - 09:25 am Toxic neuropathies in children (iatrogenous & nitrous oxide induced)
Céline Tard, Lille - France

09:25 - 09:50 am Exercise tests in muscle channelopathies
Yann Péréon, Nantes - France

09:50 - 10:15 am Motor unit number index:
A potential electrophysiological biomarker for pediatric spinal muscular atrophy
Christophe Boulay, Marseille - France

10:15 - 10:30 am Closing remarks

10:30 - 11:00 am Coffee break with partners

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Room	Oreaux Baguenaud	Pierre Sastre	Euler-Lancastria
11:00 - 11:45 am	WS4 Small fibre exam *Nancy Kuntz*	WS5 Brachial plexus *Ruth Van der Looven*	WS6 US *Amal Oulhissane-Omar*
12:00 - 12:45 pm	WS4 Small fibre exam *Nancy Kuntz*	WS5 Brachial plexus *Ruth Van der Looven*	WS6 US *Amal Oulhissane-Omar*

12:45 pm End of the 13th International Congress of Paediatric Electromyography

ORAL COMMUNICATIONS

ORAL COMMUNICATION

Justyna PIGOŃSKA[1]
Marta Banach[2]

1. EMG Laboratory, Central Clinical Hospital of Medical University Lodz,Poland
2. EMG Laboratory Collegium Jagellonian University, Cracow, Poland

Muscle ultrasound as a complementary diagnostic value in neuromuscular diseases

Ultrasonography of nerves and muscles using a high-resolution linear probe is a constantly developing field to assess their structure. It is now a complementary method to EMG in many cases.
The use of the Heckmatt scale allows a semi-quantitative assessment of the changes in the muscles examined and can be useful to indicate the muscles that are most degenerate, i.e. with potentially the greatest deviations in electromyography and indirectly in electroneugraphy.
In the current presentation, we present the case of a patient diagnosed with a mutation in the TK-2 gene (Thymidine kinase-2) , with progressive muscle paresis present. EMG showed discrete myogenic changes in the tibialis anterior muscle. Muscle ultrasound showed obliteration of muscle structure and increased hyperechogenity.

CONCLUSIONS:
Ultrasound of nerves and muscles can be a useful method to identify the most involved and therefore most characteristic lesions in a patient. The use of both methods helps to reduce the discomfort of the examination so important especially in paediatric neurophysiology.

ORAL COMMUNICATION

Hanna KÜPPER
Veronka Horber

Neuropaediatric Department, University Childrens' Hospital Tübingen

Correlation of nerve ultrasound findings with clinical and electrophysiological characteristics in Paediatric Charcot-Marie-Tooth patients

Introduction: Charcot-Marie-Tooth (CMT) neuropathy represents one of the most common hereditary neuromuscular diseases, with a prevalence of around 1:2500. It is characterised by a broad phenotypic spectrum. Published data on long-term clinical characterisation as well as electrophysiological and imaging patterns is scarce.

Methods: Within clinical care, high-resolution nerve ultrasound was conducted according to a standardized protocol in 40 Paediatric CMT patients (aged 17 months-18 years, mean 11y) with a broad range of CMT subtypes. This data was correlated to clinical characteristics (semi-standardized, according to CMT-Ped-Score, 6-minute walk test, MRC score) as well as nerve conduction study and EMG (if possible).

Results: Nerve enlargement, as depicted by nerve ultrasound (in relation to age-adjusted normal values, Druzhinin et al., 2019), was most pronounced in n=12 patients with CMT1A in a homogenous-diffuse pattern. This was already visible at a young age <2years. A teenage patient with NDRG1-related CMT4D showed a very pronounced, focally and fascicle-accentuated enlargement especially of the arm nerves, while presenting progressive loss of hand function and a severe demyelinating-axonal neuropathy with distinct denervation on EMG. By comparison, a 3 year old boy with NDRG1-related CMT4D did not show this focally enlarged pattern. N=8 patients with SH3TC2-related CMT4c as well as N=2 patients with GJB1-related CMTX and NEFL-related CMT2E demonstrated mostly normal nerve size. N=3 patient with HNPP showed typical nerve enlargement only at entrapment sites while nerve conduction also demonstrated signs of a diffuse neuropathy. A patient with FLVCR1-related HSAN (with correlating clinical-electrophysiological signs of a sensory neuropathy) showed particularly small nerves.

Conclusion: Nerve ultrasound represents a promising tool within the evaluation of Paediatric CMT. By this clinically-based examination of 40 Paediatric CMT-patients, differential nerve imaging patterns could be described, in relation to clinical-electrophysiological data.

ORAL COMMUNICATION

Peter KARACHUNSKI
Ann Van Heest, Elise Palzer, Paul J. Orchard

University of Minnesota, USA

Median nerve ultrasound as a screening tool diagnosing carpal tunnel syndrome in mucopolysaccharidosis type 1H patients

Mucopolysaccharidosis type 1H (Hurler syndrome) (MPS 1H) is a rare lysosomal storage disorder due to deficiency of α-L-iduronidase (IDUA). As a result, accumulation of substrate in multiple organs occurs, including the musculoskeletal system. Median entrapment neuropathy at the carpal tunnel is a common complication in MPS 1H. Annual screening for carpal tunnel syndrome (CTS) is a part of standard care in MPS 1H patients at the University of Minnesota. The standard approach to diagnose CTS is electroneuromyography (ENMG). This test is not well tolerated by young children, and therefore ENMG is often performed under general anesthesia. Median nerve ultrasound (MNUS) is rapid and painless. Focal enlargement of the median nerve at the carpal tunnel is diagnostic of entrapment neuropathy which is easily identifiable by MNUS.

Therefore, MNUS can be used as a diagnostic tool for identifying CTS in children with MPS 1H. In a retrospective study, we analyzed data obtained from 48 consecutive surgically naïve post bone marrow transplant patients under 10 years of age. We compared median nerve cross-sectional area (CSA) obtained by MNUS and compared with the simultaneous results from ENMG. Based on electrodiagnostic criteria we identified 10 patients with mild, 7 with moderate and 8 with severe CTS. We identified the lowest CSA threshold of 6.5 mm2 diagnostic of CTS with sensitivity of 85.3% and specificity of 93.2%. A few patients who were falsely negative had mild CTS which does not require immediate surgical correction.

We believe that MNUS shows merit in screening patients at risk for CTS, and we suggest a new paradigm for CTS screening which may include MNUS in the office. Patients who met the threshold of CSA for diagnosis of CTS should undergo confirmatory testing with ENMG. Use of MNUS may reduce frequency of anesthesia used for ENMG in these patients.

ORAL COMMUNICATION

Arnaud CHEFDOR
Armelle Magot, Paul Arnolfo

CHU Nantes, France

Carpal tunnel syndrome in pediatric patients : Presentation of Two Cases, One Typical and One Atypical, Associated with PIK3 Pathway Abnormalities, and review of the literature

Carpal tunnel syndrome (CTS) is a rare condition in children. The most common causes are the mucopolysaccaridoses and mucolipidoses diseases. Recently, variants of the PIK3 pathway have been described in congenital malformation, which is another cause of CTS. Patients and methods: We report two cases of 11 and 12 year-old boys recently diagnosed with genetically proved PIK3CA (for the first patient) and PIK3R1 (for the second) mutations. For the first patient electrophysiological studies (EDX) showed an isolated left CTS with secondary motor and sensory axonal loss. The EDX of the second patient showed chronic denervation in abductor pollicis brevis associated with unusual sensory-motor axonal multineuropathy. Ultrasonography (US) was performed for the 1st patient and showed lipomatosis of nerve. For the 2nd patient US and magnetic resonance imaging were performed and found an aspect of infiltration of the nerve fascicles compatible with a tumoral aspect. Discussion and conclusions: The 1st patient illustrates a typical lipomatosis of nerve integrating into CLOVES syndrome. Lipomatosis of nerve has pathognomonic US and MRI features. For the 2nd patient, the hypothesis of the nerves containing increased amount of endoneurial connective tissue and interstitial fat, could be proposed. The EDX pattern of the 2nd patient was similar to those encountered in chronic entrapment neuropathies.

We report two cases of CTS in children with alteration of the PIK3 pathway but with different features clinical-radiological presentations: a classical lipomatosis of nerve for one and atypical for the second.

ORAL COMMUNICATION

Laura NASTASI[1]
Gerald Cooray[1], Amjad Aldrees[1,2], Sheba Azam[1], Jacquie Deeb[3]

1. Department of Neurophysiology, Great Ormond Street Hospital, London, United Kingdom
2. Division of Neurology, Department of Paediatrics, King Abdullah Specialised Children's Hospital, Riyadh, Saudi Arabia
3. Department of Neurophysiology, Queen's Hospital, Romford, United Kingdom

ENMG in paediatric ICU

Background: Neuromuscular disorders encountered in neonatal and paediatric intensive care units (ICU) include a broad spectrum of both acute and chronic pathologies, alongside neuromuscular complications of prolonged ICU stay. Electroneuromyography (ENMG) plays an important role in facilitating a timely diagnosis and management of these children. This study aims to share our experience in a tertiary paediatric centre over a decade.

Methods: We reviewed the EMG studies performed for evaluation of neuromuscular conditions in children staying in the intensive care unit at Great Ormond Street Hospital in London from 2010 to 2019. Electrodiagnostic data was extracted from departmental database. Z scores were calculated for each ENMG parameter based on e-norm reference values. Patients' medical records were reviewed, and clinical diagnoses were identified based on clinical profile, laboratory, genetic, imaging and pathology test results.

Results: A total of 431 EMG tests were performed in 351 patients (mean age 42.2 months), 57% of which were infants. Isolated respiratory distress was the most frequent reason for referral. Most patients underwent routine nerve conduction and electromyography, while some required specialized tests like diaphragmatic-EMG and stimulation single-fibre EMG. We classified the electrodiagnostic findings into motor neuronopathy (n=92), myopathy (n=49), neuropathy (n=45), neuromuscular junction disorder (n=31), critical illness neuromyopathy (CINM) (n=28), other (n=4) and normal (n=102). A positive electrophysiological-clinical correlation was found in almost all cases within the CINM and neuropathy group; correlation ranged between 50% and 70% within the remaining groups. A wide range of pathologies were identified in each group.
Conclusions: This study confirms the valuable role of ENMG as an adjunctive tool in the management of complex neuromuscular conditions in the paediatric intensive care unit and highlights the most commonly encountered diseases in this setting and electrophysiological characteristic in this patient group.

Acknowledgments: Dr Matthew Pitt, Mr Daniyal Motan, Miss Kitty Howse

ORAL COMMUNICATION

Leena LAURONEN[1]
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1. Department of Clinical Neurophysiology, Children's Hospital, HUH Medical Imaging Center, University of Helsinki and Helsinki University Hospital, Helsinki, Finland
2. Department of Child Neurology, Children's Hospital, Pediatric Research Center, University of Helsinki and Helsinki University Hospital (HUH), Helsinki, Finland

Polyneuropathy in vincristine treated pediatric ALL patients

Objective: To characterize the occurrence and development of vincristine-induced neuropathy (VIPN) in the early treatment of children with acute lymphoblastic leukemia (ALL).

Methods: This prospective study comprised of 35 2-14-year-old ALL patients receiving vincristine. We performed systematic clinical and electrophysiological studies at both the time of diagnosis and on one to two time points during the first months of treatment.

Results: After vincristine treatment, all patients had axonal sensorimotor PNP in ENMG. In 34/35 patients, the motor and in 24/35 the sensory responses were decreased. However, some children had no PNP symptoms despite moderate ENMG findings, and not all clinical symptoms were correlated with abnormal ENMG. Interestingly, in 3 patients PNP was most prominent in the upper limb.

Conclusions: Pediatric ALL patients treated with vincristine are at marked risk of developing VIPN. Pediatric VIPN is a sensorimotor, predominantly motor axonal neuropathy. VIPN is difficult to detect by symptoms and clinical examination only, but can be detected even in its early phase by ENMG.

ORAL COMMUNICATION

Julie PIARROUX[1]
Evgenia Dimopoulou[1], Guillaume Taieb[2], Sarah Souvannanorath[3], Emmanuel Roze[4], Laurence Lion-François[5], Marie-Aude Spitz[6], Emmanuel Broussolle[7,8,9], Chloé Laurencin[8,10], Jean-Baptiste Chanson[11], Johanna Belleville-Goffeney[12], Pierre Meyer[1], Mirna Khalil[13], Maelle Dereure[14], Diane Doummar[15], Hugues Chevassus[13], Emmanuelle Apartis[17]*, Agathe Roubertie[1,18]*

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**: Contributed equally to this work.*

Electrophysiological characterization of essential tremor in children and adolescents

Objective: Essential tremor (ET) is considered the most frequent abnormal movement in the general population, with childhood onset in 5 to 30% of the patients. This study aimed to characterize the clinical and electrophysiological features and functional impact of ET in children and adolescents. Here we will focus on the electrophysiological findings.

Methods: A multicenter, descriptive cross-sectional study enrolled patients 18 years with a definite diagnosis of ET according to the International Parkinson and Movement Disorders Society criteria. Data concerning sex, family history, clinical and electrophysiological characteristics of the tremor, neurological examination and impact on quality of life according to validated scales were collected.

Results: 9 male and 9 female patients were included (mean age of 13.9 years). Tremor was characterized by: upper limb onset at a mean age of 6.5 years; kinetic tremor in all of the patients combined with postural tremor in 17 and rest tremor in 3; tremor mean frequency of 7.6 Hz, mean burst duration of 82.7 ms; in two patients, tremor was very subtle and could not be recorded. There was no difference in tremor frequency between boys and girls, nor according to age at onset, age at examination, duration of disease or the presence of family prior. We identified mild myoclonic jerks on the polymyographic recordings in 7 patients.

Discussion: Electromyographic characteristics of ET in children are close to those found in older population, with a few interesting specificities: amplitude can be low, making the tremor difficult to record, and tremor seems to be associated with a mild myoclonus in some children.

POSTERS

POSTER - P1

Yvan DE FERAUDY[1]
Aleksandra Nadaj-Pakleza[2], Vincent Laugel[1], Jean-Baptiste Chanson[2]

1. Neuropediatrics Department, Hautepierre University Hospital, Strasbourg
2. Electrophysiology Laboratory, Department of Neurology, Hautepierre University Hospital, Strasbourg

Utility and limitations of electroneuromyography in pediatrics: A study cohort of 137 patients

Electroneuromyography (ENMG) is a commonly used diagnostic tool for neuromuscular pathologies. However, this technique may cause discomfort in young patients, so its appropriateness should be thoroughly considered in pediatrics. Our objective was to assess its clinical utility in children of different age groups, especially the prevalence and predictive factors of ENMG abnormalities, and the agreement between ENMG findings and final diagnoses.

We included all patients under 16 years of age who underwent ENMG at the University Hospital of Strasbourg between 2017 and 2021. Clinical and ENMG data were collected retrospectively.

Among the 137 included patients, 41 (30%) had an abnormal ENMG. The highest proportion of abnormal ENMGs was observed before the age of 1 (50%). The most frequent ENMG abnormality was myopathic pattern in needle analysis in this group while a diffuse neuropathy was more commonly found in older patients. The presence of abolition/reduction of osteotendinous reflexes, muscle weakness, paresthesias, and/or hypoesthesia (excluding neuropathic pain) increased the likelihood of an abnormal ENMG ($p < 0.05$). The concordance between ENMG results and final diagnosis was stronger for neuropathic findings (95%) than for myopathic abnormalities (44%). Turns-amplitude analysis was more sensitive, and the presence of excessively early and rich recruitment was more specific to detect myopathic diseases. Notably, ENMG failed to detect the single case of congenital myasthenia included in the study. All instances of false negatives and false positives occurred in patients under 5 years old, mainly before the age of 1.

ENMG remains a crucial diagnostic tool in pediatric medicine, demonstrating relatively good agreement with final diagnosesespecially to diagnose a neuropathy in children of more than 5 years of age. ENMG is also useful to detect potential myopathies in children of less than 1 year of age but the agreement with final diagnosis is lower.

POSTER - P2

Amanda SOEBADI[1]
Amanda Soebadi[2], Cut Nurul Hafifah[2], Susyana Tamin[2], Elvie Zulka Kautzia[2], Luh Karunia Wahyuni[2], Rizky Kusuma Wardhani[2], Madeleine Ramdhani Jasin[2], Fitri Primacakti[2], Dina Muktiarti[2]

1. *Neurology Division, Department of Child Health, Universitas Indonesia Faculty of Medicine*
2. *Nutrition and Metabolic Disease Division, Hemato-oncology Division, Allergy-immunology Division, Respirology Division, Department of Child Health; Department of Physical Medicine and Rehabilitation, Universitas Indonesia Faculty of Medicine*

The mysterious case of a teenage girl with myopathy, peripheral neuropathy, and unintentional weight loss

A 15-year-old girl with non-consanguineous parents presented with weakness of all four extremities since three months prior. She had difficulty squatting and rising from seated on the floor. She also complained of a "heavy" feeling when lifting her arms overhead. She had difficulty swallowing and coughing, leading to frequent choking. Over the next three months the weakness remained relatively unchanged. Since nine months prior she also lost weight from 85 kg to 50 kg. There were no sensory or autonomic symptoms. Examination showed symmetric, proximal-dominant weakness in all four extremities with scapular winging. Deep tendon reflexes were normal in the lower and reduced in the upper extremities; pathologic reflexes were absent. There was atrophy of the deltoids, triceps, biceps, hamstrings, and glutei, with relative sparing of the rectus femoris and gastrocnemius. Spinal deformity was absent. Serum creatine kinase was 5363 U/L; other laboratory parameters were unremarkable. Echocardiography was normal. Nerve conduction studies showed reduced CMAPs in both peroneal nerves on proximal and distal stimulation, with normal velocity and latency. Tibial, median, and ulnar nerve conduction studies were within normal limits. Needle EMG of the anterior tibial and deltoid muscles showed increased spontaneous activity, normal recruitment and activation, and mildly polyphasic MUPs with increased amplitude but normal duration. Endoscopic and fluoroscopic swallow studies revealed oropharyngeal-phase neurogenic dysphagia with a penetration-aspiration score of 7, and weakness of the plica vocalis and epiglottis. Whole exome sequencing revealed a COL6A2 missense mutation, known to be associated with Bethlem myopathy, as a variant of uncertain significance. She felt subjective improvement with prednisone given on a 7-days-on, 7-days-off basis; she is currently in a prednisone taper. She has started to gain weight after the introduction of tube feeding. Spirometry was within normal limits. Since her genetic testing results only partially explain her symptoms, further diagnostic efforts are underway, including testing for myositis-specific antibodies.

POSTER - P3

Thouraya BEN YOUNES
Ichraf Kraoua, Zouhour Miladi, Abir Zioudi, Hedia Klaa, Hanene Benrhouma, Ilhem Ben Youssef Turki

LR18SP04 and Pediatric Neurology department, National Institute Mongi Ben Hmida of Neurology, Tunis, Tunisia

Contribution of ENMG to the diagnosis of neurometabolic diseases

Introduction: Inherited metabolic disorders (IMD) constitute a heterogeneous group of rare genetic conditions. Peripheral nervous system involvement is common. The contribution of electroneuromyogram (ENMG) in the diagnostic approach of these pathologies is crucial but rarely studied.

Objective: Report the characteristics of peripheral neuropathies (PN) during IMD in children and discuss the contribution of ENMG in the diagnostic approach of these diseases.
Materials and methods :
Retrospective descriptive study over a period of 17 years (from 2005 to 2022), including patients followed in the pediatric neurology department at the National Institute Mongi Ben Hmida of Neurology in Tunis, for IMD with PN. Clinical and electrophysiological data were evaluated.

Results: We collected 73 patients. PN revealed IMD in 10 cases. The average age of onset of PN was 4.8 years. The PN were most often diffuse polyneuropathy (69/73). Carpal tunnel syndrome was found 2 patients with mucopolysaccharidosis. Demyelinating neuropathies were predominant (40/73) (metachromatic leukodystrophy (23 cases), Krabbe leukodystrophy (6 cases), mitochondriopathy (7 cases), Congenital-Disorder of Glycosylation (1 case), vitamine E deficiency (1 case), Nieman pick type C (1 case), fumarase deficiency (1 case). Axonal neuropathies were found in 25 cases (mitochondriopathy (16/25), GM2 gangliosidosis (2/25), Fabry disease (2/25), Congenital-Disorder of Glycosylation (1/25), Farber disease (1/25), Biotinidase deficiency (1/25), hyperglycinemia without ketosis (1/25), acute intermittent porphyria (1/25). The mixed mechanism was found in 8/73. There were sensorimotor involvement in 55/73, pure sensory involvement in 13/73 and pure motor involvement in 5/73.

Conclusion: Peripheral neuropathies are common in IMD and can guide our diagnostic approach. The ENMG must be part of the etiological investigations in cases of suspicion of IMD. Early diagnosis allows appropriate treatment to be initiated in patients with a treatable condition.

POSTER – P4

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The motor unit number index (MUNIX) in control children: Reference values and reliability

Introduction: The MUNIX (Motor Unit Number IndeX) is a technique that estimates the number of motor units (MUNIX) and their size (MUSIX, Muscle Size IndeX) in a given muscle. This technique is based on the recording of the supramaximal compound muscle action potential (CMAP) and surface electromyography at different levels of voluntary muscle contractions. In healthy adult, the MUNIX technique has been shown to be an useful and reliable electrophysiological biomarker and reference values have been determined. There are very few studies on the use of MUNIX in paediatrics (small sample size). It is therefore necessary to assess the MUNIX technique in pediatrics on larger samples, in particular in a population of control children, to provide reference values as a function of advancing age and reliability.

Methods: Sixty control children (28 females;median(Med) age 10 years [interquartile range(IQR),4-14 years]) were enrolled. The sample is divided into subgroups according to age distribution: under 5 years (n1 =16), between 5-10 years (n2 =15), then 10-15 years (n3 =21), and 15-17 years (n4 =9). The MUNIX technique was evaluated in the abductor digiti minimi (ADM) and abductor pollicis brevis (APB).

Results: APB MUNIX increased between n1 and n3 (Med(IQR) 118(87-155) vs. 203(113-286)), but for ADM this increase occurred earlier, between n1 and n2 (Med(IQR) 118(117-131) vs. 195(155-220)). The intra-rater class coefficient correlations (ICC>0.75), for experienced operator, were interpreted as good reproducibility of all measures (MUNIX,CMAP,MUSIX) in all subgroups. But the inter-rater ICC for experienced vs non-experienced operators were not interpreted as good reproducibility (<0.75) of all measures in n1 and n2.

Discussion: This study provided reference values for the MUNIX technique in a population of control children and assessed its good reproducibility for experienced operators. The MUNIX technique was shown to be a potential biomarker for disease progression and treatment response.

POSTER - P5

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Improved gait and radiological measurements after injection of botulinum toxin into peroneus longus in young children with Unilateral Spastic Cerebral Palsy (USCP) and equinovalgus gait

Background: Children with cerebral palsy develop foot deformities due to a combination of factors including muscle shortening, hypertonia, weakness, and cocontraction of muscles acting at the ankle joint resulting in an altered gait pattern. We hypothesized these factors affect the peroneus longus (PL) and tibialis anterior (TA) muscles couple in children who develop equinovalgus gait first followed by planovalgus foot deformities. Our aim was to evaluate the effects of abobotulinum toxin A injection to the PL muscle, in a cohort of children with unilateral spastic cerebral palsy and equinovalgus gait.

Methods: This was a prospective cohort study. The children were examined within 12 months before and after injection to their PL muscle. Twenty-five children of mean age 3.4 (S.D.: 1.1) years were recruited.

Results: We found significant improvement in foot radiology measures. Passive extensibility of the triceps surae did not change, whereas active dorsiflexion increased significantly. Nondimensional walking speed increased by 0.1 (95% confidence interval [CI], [0.07, 0.16]; $P < 0.001$), and the Edinburgh visual gait score improved by 2.8 (95% CI, [-4.06, -1.46]; $P < 0.001$). Electromyography showed increased recruitment for gastrocnemius medialis (GM) and TA but not for PL during the reference exercises (standing on tip toes for GM/PL, active dorsiflexion for TA) and decreased activation percentages for PL/GM and TA across sub-phases of gait.

Conclusions: One key advantage of treating the PL muscle only might be to address foot deformities without interfering with the main plantar flexors that are instrumental to support body weight during gait.

CONGRESS DINNER

NOVEMBER 14TH, 2023 | 7:00PM

Venue l'Escal'Atlantique
Submarine pen in Saint Nazaire

7:00pm Bus departure
from the Palais des Congrès
La Baule

With the participation
of the improvisation troupe La LINA

SF-ENMG
Société Francophone d'ElectroNeuroMyoGraphie

Filnemus
Filière Neuromusculaire

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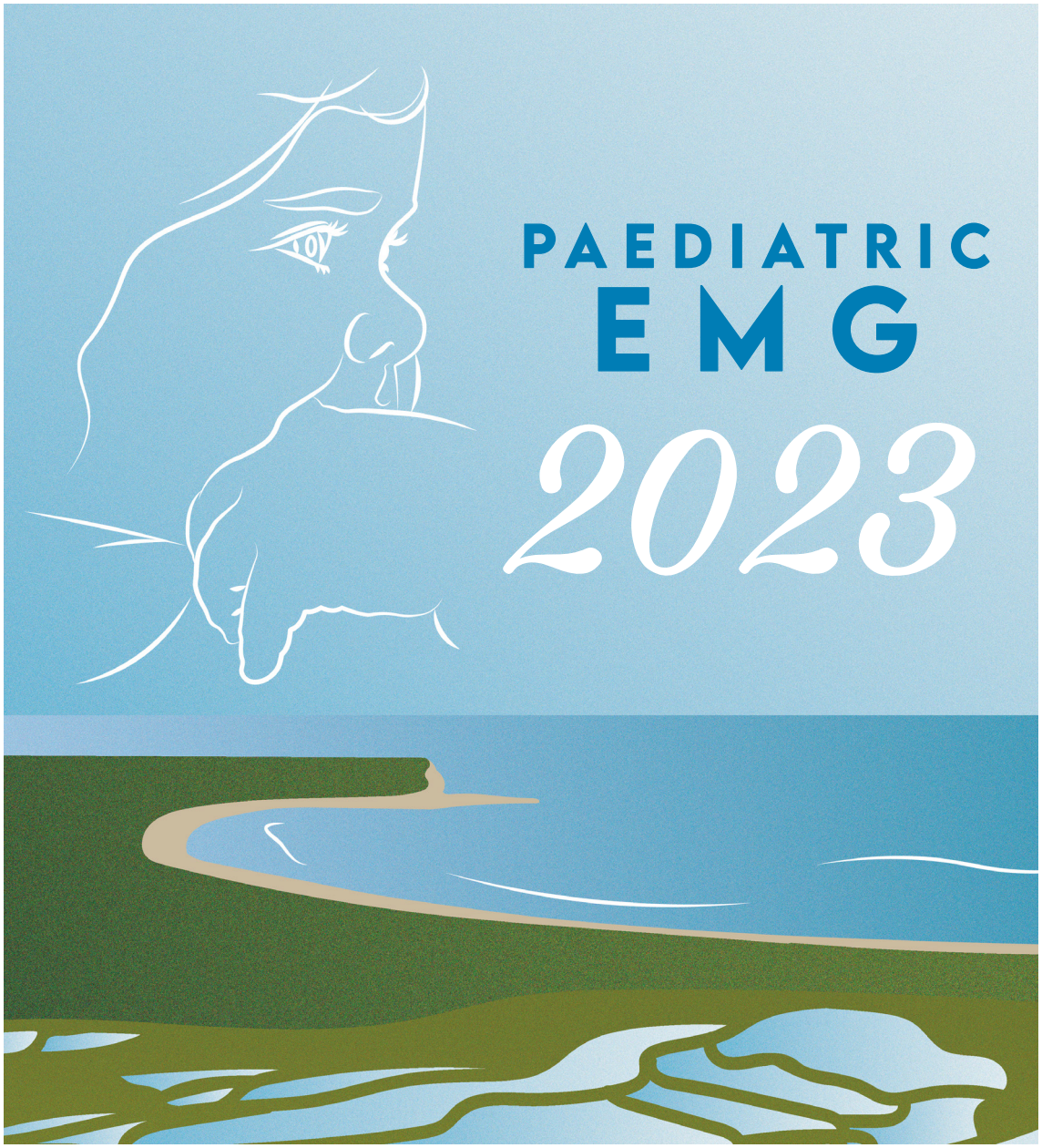
LIGUE D'IMPROVISATION NANTES ATLANTIQUE



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