Results: Out of 398 enrolled patients (215F, 183M, mean age 41.28 ± 15.68), 48.40% had IIH; 26.58% had migraine (including a few cases of migraine with aura), 20.85% had tension-type headache, a few patients had other types of headache. PIH was observed in 23.36% of patients (93), occurring pre-ictally in 26 cases, ictally in 3 and post-ictally in 75 (11 patients had different types of PIH). Out of 105 patients with interictal migraine, 47.61% had PIH (p = 0.0001), that was pre-ictal in 17.14% (p < 0.0001) and post-ictal in 39.04% (p = 0.0001). There were no significant associations with the epilepsy syndrome (generalized or focal). Post-ictal headache had migrainous features in 50.66% of patients and tension-type headache-like in 40.00%. The occurrence of post-ictal headache was significantly associated with antiepileptic drug polytherapy (p < 0.0001), high seizures’ frequency (p = 0.001) and the presence of IIH (p < 0.0001).

Conclusion: These data show that migraine is the most represented type of headache in our patients. Subjects with interictal migraine resulted more prone to develop both pre-ictal and post-ictal headache. The occurrence of the latter is higher in patients with polytherapy, high seizures’ frequency and coexistence of IIH.

p729
TWO CASES OF CHOREA-ACANTHOCYTOSIS WITH EPILEPSY
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Purpose: We aimed to present two cases of Chorea-acanthocytosis (ChAc) whose initial manifestations were epileptic seizures. Case 1: A 42-year old woman with newly onset unprovoked seizures admitted to our clinic and diagnosed as epilepsy with complex partial secondarily generalized seizures. Electroencephalography (EEG) revealed right frontocentral spikes and occipitoparietal slow waves. Cerebral magnetic resonance imaging (MRI) showed bilateral caudate nucleus atrophy and acanthocytes were detected at blood smear. She was diagnosed as ChAc. Case 2: A 39-year old woman admitted to our clinic with involuntary movements and gait disorder. Neurologic examination revealed chorea, bizarre gait, involuntary oral biting and tics. She had a history of recurrent seizures and was taking carbamazepine for one year. Seizures were complex partial secondarily generalized and EEG revealed right temporal spikes. Cerebral MRI showed bilateral caudate nucleus atrophy and acanthocytes were detected at blood smear. She was also diagnosed as ChAc.

Conclusion: Seizures may be the initial manifestation of ChAc. It should be kept in mind in adult onset epilepsy patients with movement disorders like chorea, tics, unstable or bizarre gait and oral mutilation.

p730
NATIONAL REGISTRY OF DRAVET’S SYNDROME AND OTHER SYNDROMES CORRELATED WITH GENES SCN1A AND PCDH19
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Purpose: Epidemiological surveillance of Rare Diseases through Patient Registers has been recognized as one of the priorities in the Public-Health-Programme strategic intervention of the European Commission. Dravet Syndrome (DS), also known as Severe Myoclonic Epilepsy of Infancy, is a rare epileptic encephalopathy with onset in the first year of life with recurrent epileptic seizures, precipitated by fever, and progressive neurodevelopmental disability in the following years. Some clinical features of DS are shared by other rare syndromes due to SCN1A or PCDH19 genes mutations. In order to develop new therapeutic solutions and social-health services to improve patients’ quality of life, the “Dravet Italia” – a Non-profit Association (http://dravetitalia.org/) – promoted the development of the Italian Registry of DS and other Syndromes related with SCN1A or PCDH19 genes mutations (ReSiDraS).

Method: The ReSiDraS has been developed by a multifaceted working group consisting of expert clinicians (www.dravetitalia.org/it/comitato-medicodiscientifico), representatives of Patient Associations, and experts in Disease Registries, as well as Information Technologies specialists by the FTGM-CNR (www.ftgm.it). The working group, after careful data analysis, has identified the data needed and the registry structure. The FTGM implemented the registry in a web-based format, accessible through authentication of certified users via username and password. Data will be gathered by users after receiving the patients’ parents informed consent.

Results: The web-based ReSiDraS registry is now ready and the beta-version is currently under evaluation in order to remove any possible bug before the official release. We aim to start collecting data from the entire Italian territory by the end of 2014, reaching a full coverage within few years.

Conclusion: The data collected by ReSiDraS will allow us a better understanding of the clinical, genetic, and epidemiological aspects of the different diseases included, possibly shedding light on genotype-phenotype correlations, drugs currently used and their efficacy, comorbidities, and long-term outcome.

p731
EPILEPTIC SEIZURES IN PEDIATRIC POPULATION WITH BACTERIAL MENINGITIS IN A DEVELOPING COUNTRY
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Purpose: To evaluate occurrence and relevance of seizures in children with community-acquired bacterial meningitis in a public secondary care university hospital in Sao Paulo, Brazil.

Method: A retrospective study in which clinical data of pediatric inpatients with community-acquired bacterial meningitis was reviewed in a 10-year period (2003–2013).

Results: 101 children (70% male), median age 28 months. (IQR 7–69); epileptic seizures occurred in 10 (9.9%). Patients with seizures were younger (median age 15 months [IQR 7–19]) vs. 35 months [IQR 7–74]; p = 0.01), had higher median serum leukocyte count (18.5 × 109/l [IQR 17.3–26.8] vs. 15.8 × 109/L; p < 0.001), had higher median CSF leukocyte count (3.498 × 8.000) vs. 672/µL [IQR 161–2345]; p = 0.03) and higher median CSF protein levels (257 mg/dl [IQR 220–288] vs. 69 mg/dl [IQR 43–157]; p < 0.001) than patients without seizures. From 10 patients with seizures, in 4 pneumococcus (all these cases occurred before implementing free public pneumococcus immunization for infants in 2010 in the country) was isolated in CSF, in 3, meningococcus, in one, type-A Haemophilus and in two, no agent was identified. Neuroimaging was done in 42% of patients and in all episodes with seizures, revealing a focal lesion in 8% of patients, only one who had seizures. EEG was performed in all seizure episodes and was abnormal in 60% showing focal (30%) or diffuse slowing (30%). One patient had interictal epileptiform activity in frontal regions. Antiepileptic drugs
National Registry of Dravet’s Syndrome and other Syndromes correlated with genes SCN1A and PCDH19

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**INTRODUCTION**

Dravet Syndrome (DS), also known as severe infant myoclonic epilepsy, is a rare form of epilepsy associated with disturbances of neurological development. Incidence is estimated at between 1/20,000 and 1/40,000, with a greater frequency in males than in females. The incidence of the disease has been estimated at between 1/20,000 and 1/40,000, with a greater frequency in males than in females; symptoms appear in the first year of life with recurrent epileptic crises, worsened by fever. There is an ample spectrum of clinical variability associated with genetic alterations on SCN1A.

Patient registers have been recognized by the European Community as one of the priorities of strategic intervention in the Rare Illnesses sector. (1) These constitute an essential instrument for improving understanding of the illness through the systematic and continual registration of data for basic research, clinical and epidemiological, with the aim of developing new therapeutic solutions and the programming of social-health services to improve patients’ quality of life and that of their families. The possibility of having trustworthy patient data regarding the entire national territory is a fundamental element for understanding phenomena linked to the distribution in space and in time of the illness. The Dravet Italy Non-profit Association (http://dravetitalia.org/) has promoted the development of the “National Registry of Dravet Syndrome and other Syndromes correlated with mutations on genes SCN1A and PCDH19” in collaboration with the Medical Scientific Committee (www.dravetitalia.org/it/comitato-medico-scientifico) and with FTGM (www.ftgm.it). This organizes the development of the computerized registry accessible via web only by centers authorized and validated by the Committee.

The Registry model has been developed by a working group consisting of expert clinicians, members of the Scientific Medical Committee (fig. 1), representatives of patient associations, experts in illness registries and information technologies useful for their implementation. The working group, after having identified the main aims of the Registry, elaborated its structure, establishing 9 headings: Anagraphic Data; Genetic Investigations; Family History (Familial Anamnesis); Personal History (Personal Anamnesis); Onset of Epileptic Crises; Crises Follow-up; Neurological Follow-up; Therapy; Adverse events. Each of these headings is composed of a number of variables, obligatory and optional, to be inserted. In the pilot phase the register’s data will be gathered and inserted by specialized medical personnel attached to medical structures identified by the Association (fig. 2). Insertion of data will be through access to the web site of the Registry, subject to verification by the Medical Scientific Committee and successive activation of the user by the administrators of the site (FTGM) who will create a log-in and send an e-mail to the user with instructions for the initial access and the memorization of his/her personal password. Data will be gathered after having consigned to the patient the relative informative documentation and having obtained his/her acceptance to the use of the data. Identification of the patient will be realized through the generation of a code encrypted at the moment of the insertion of the surname, name, date of birth and place of birth (or fiscal code). The computerized administration of the Registry will determine the modalities of memorization, of transmission and of access to the electronic data in accordance with the relative legal norms, in particular in accord with the Articles of Attachment B of D.lgs. 30 giugno 2003 n. 196 “Codice in materia di protezione dei dati personali” (“Code on Personal Data Protection”), and successive modifications and integrations. The protocol in its entirety and the relative modalities of data treatment has been submitted to the ethic committee of reference. (specify) The administration of the register will be entrusted to a Coordinating Committee formed by the Medical Scientific Committee, the Dravet Non-profit Association Italy representing the families, and by the FTGM.

**EXPECTED RESULTS**

- Acquisition of clinical, genetic and epidemiological data regarding the illness and its correlated syndromes
- Evaluation pharmacological treatments used and their efficacy
- Evaluation outcome neurological, epileptological and behavioural cognitive
- Evaluation complications and/or comorbidity associated
- Possibility of conducting studies of correlation between genotypes and phenotypes

**CONCLUSIONS**

The Registry could represent an important instrument for the systematization of data regarding DS and correlated syndromes, in order to improve understanding of the illness and the knowledge of the related research. Reports and up-to-date graphics of added data which could be essential to promote the knowledge of the illness and its social and economic impact will be made available to all interested parties, in the hope of promoting and sustaining scientific research, with the aim of discovering innovative therapeutic options for the management of DS and correlated syndromes.

**BIBLIOGRAPHY**