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FOREWORD

Georges Dassis
President
European Economic and Social Committee (EESC)

Once upon a time those wishing to know the future could consult the Oracle of Delphi. This High Priestess must have disappointed or baffled her "customers" on more than one occasion: sometimes she did not reply at all, or her answer was rather – cryptic. But the customer had been warned: inscribed on the Temple of Apollo was not "Here stands the god of whom you may ask all you would like to know", but rather "Γνῶθι σεαυτόν". Know thyself. Socrates ultimately saw this as a way of saying "try to be reasonable" (σωφρονεῖν).

It would seem that certain people have little knowledge of themselves, or of others. Unless some secret plan has passed me by, I wonder at any rate whether these people are reasonable: 2016 saw two of Europe's prime ministers resign the day after failure in a referendum which they themselves had recklessly launched.

In the first case – the United Kingdom – I did not risk predicting the result itself, but I did dare to predict the damage it would do in perpetuating a divide in the population, particularly in regional terms, rather than seeking to repair it. I'm still wondering how one can venture to ask the people's opinion without knowing what could happen, or what the legal consequences could be, if their response proved not to be what one hoped.

"I do not want to make any predictions for 2017, but I do hope that the European Union will finally start to regain a sense of its values as laid down in the first articles of its Treaty, that men and women will come forward to defend those values in tangible ways, and that citizens will have reasons to regain hope in something solid, intelligent, generous, peaceful and reasonable."

In the second case – Italy – the people were asked a set of questions whose ultimate aim was to reduce the number of instruments of representative democracy, including participatory democracy, since the intention was to abolish the National Council for Economics and Labour. Paradoxically, we then saw populists amongst those defending the institutions of the good old constitution, established by democratic parties from all sides in the wake of the horrors of fascism and war.

What I find deeply troubling is the ever-increasing legitimisation of excesses, attitudes lacking in nuance, verbal provocations, absurd promises and the impression of chaos which all this creates. I fear that the various forms which demagogy can take will leave their mark over everything, causing confusion in the population and leading to the most odious exploits.

The year 2016 also saw – in France, Belgium and Germany – crimes committed by a handful of fanatics who murdered and tortured at random in "civil society", the society represented by the Committee of which I am

president. There are not many individuals capable of committing this type of atrocity, but they are horrifyingly dangerous. I especially want them not to win in any way. Any way at all. We must track them down, but we must not taint our own lives with permanent fear nor aspire to turn our society into one of mass control. Nor can I accept that the contemptible acts of a few madmen should legitimise the rejection of hundreds of thousands of innocent and peaceful people. It makes me furious to hear the desperate, cynical and nihilistic arguments of future extreme-right voters: "When they're in power, I will suffer. My situation, my own rights, especially social entitlements, will deteriorate, and I know it. But immigrants and refugees, who are taking advantage of welfare, will suffer even more, and that's what counts".

Humankind's worst enemy, the thing holding us back, is jealousy. And yet there is a way to overcome this vice: first of all we can welcome the happiness of others and be happy about it ourselves. Next, solidarity, the core principle of trade unionism and the EU – to name but two things particularly close to my heart – is not unpleasant or even overly expensive in relation to the benefits it brings.

I do not want to make any predictions for 2017, but I do hope that the European Union will finally start to regain a sense of its values as laid down in the first articles of its Treaty, that men and women will come forward to defend those values in tangible ways, and that citizens will have reasons to regain hope in something solid, intelligent, generous, peaceful and reasonable. The key to our progress can be found in the mottoes of the three neighbouring countries recently hit by terrorism: liberty, equality, fraternity, unity and justice. ■

European Economic and Social Committee

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WELCOME

Welcome to the February edition of Adjacent Government. As it is the first one of 2017, I would like to wish all our readers and contributors a very Happy New Year. 2017 has started with a bang, with the inauguration of Donald Trump in the US and the Supreme Court ruling that the UK government must consult Parliament before triggering Article 50. Change is also afoot in the European Parliament, where Antonio Tajani has been named as the new President.

This issue starts with a foreword from President of the European Economic and Social Committee (EESC), Georges Dassis. In his opening article, Dassis gives thought to events of last year and looks forward to how we can progress this year.

The health section starts by highlighting World Cancer Day 2017, with a special focus headed up by EU Commissioner for Health, Vytenis Andriukaitis. The Commissioner's article stresses the need for prevention and joint efforts to tackle rare forms of cancer. Adjacent Government also interviews Dr Percy Ivy from the National Cancer Institute at the National Institutes of Health in the US. She sheds light on clinical trials and their integral role in cancer research. The focus also features articles from SIOPE, Cancon and Cancer Research UK.

Our extensive health section highlights a number of key healthcare challenges, including obesity, with articles from RCPCH, WHO Europe and the Canadian Institute of Health Research. HIV/Aids in Sweden, low vision, innovative medicines, and smart hospitals are also discussed.

Another area we give focus to in the February edition is wildlife crime prevention in the environment section. EU Commissioner for Environment Karmenu Vella and MEP Catherine Bearder both highlight this problem. "We need tougher action on wildlife trafficking', states Bearder in her informative piece, and Commissioner Vella highlights the EU Action Plan against Wildlife Trafficking.

In the environment section we also look at polar research, in the Arctic regions of Canada, as well as Antarctica. An article from Polar Knowledge Canada outlines how they bring together indigenous and scientific expertise to look at issues faced by Arctic communities today.

Other topics within the publication include: Erasmus+, Hungarian rail infrastructure, renewable energy sources in Canada, Digital Society, and cyber security in healthcare.

Elsewhere in this winter issue, we shine the light on a number of key HR topics, including women in leadership, HR compliance and retaining talent. Articles come from such key organisations as CIPD, Acas and PPMA to outline these areas.

As always I hope you find the February edition informative and useful, and I welcome any feedback you may have.

Laura Evans
Editor
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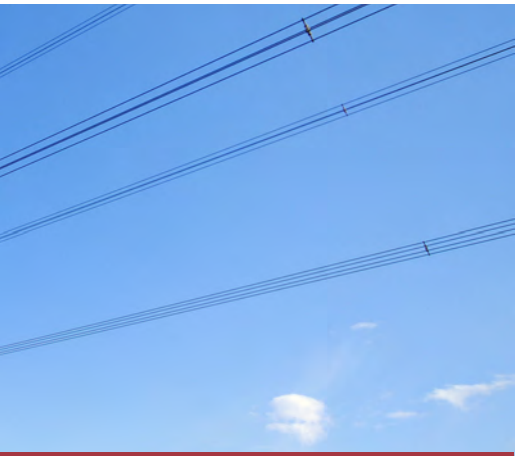

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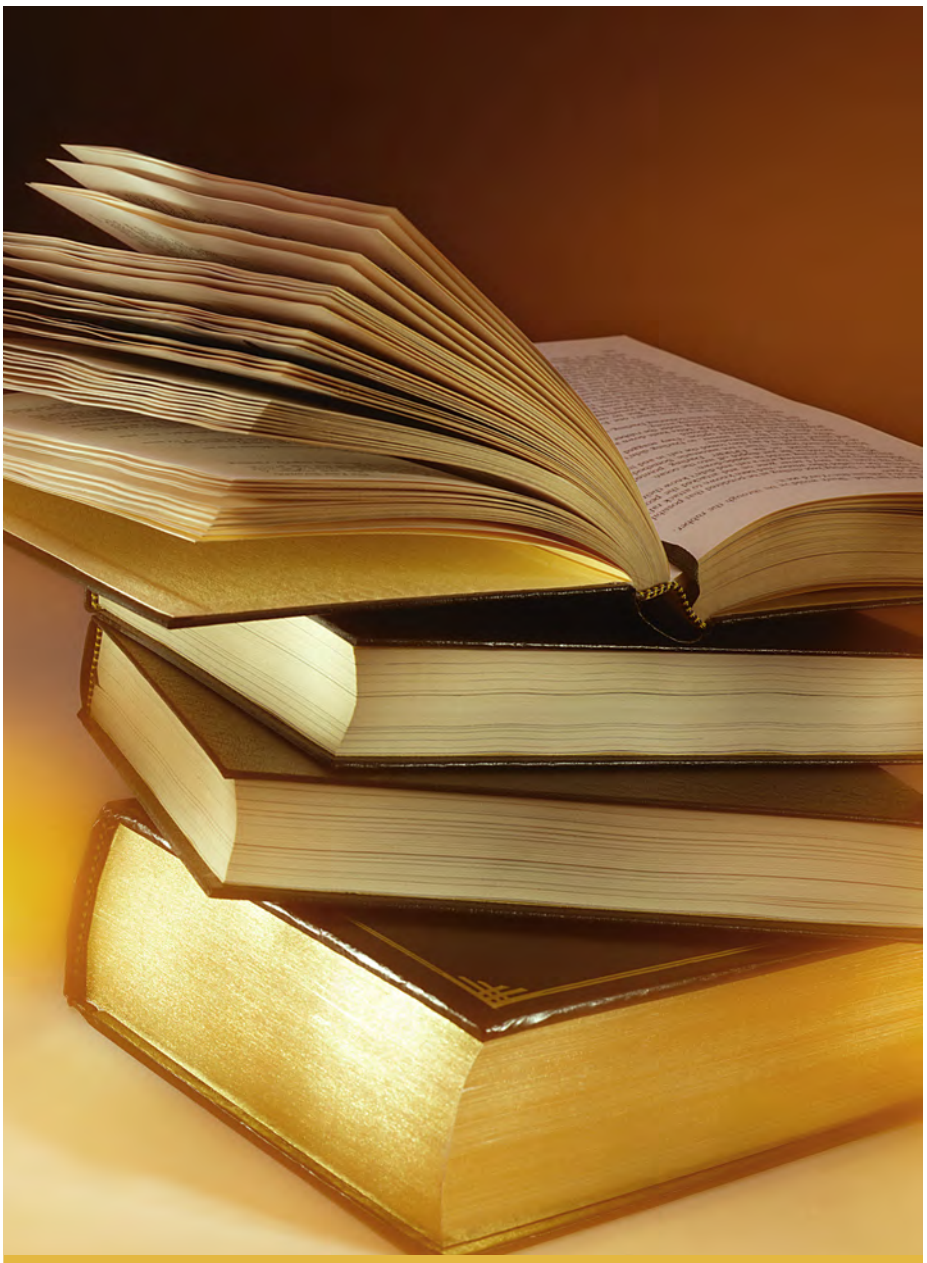

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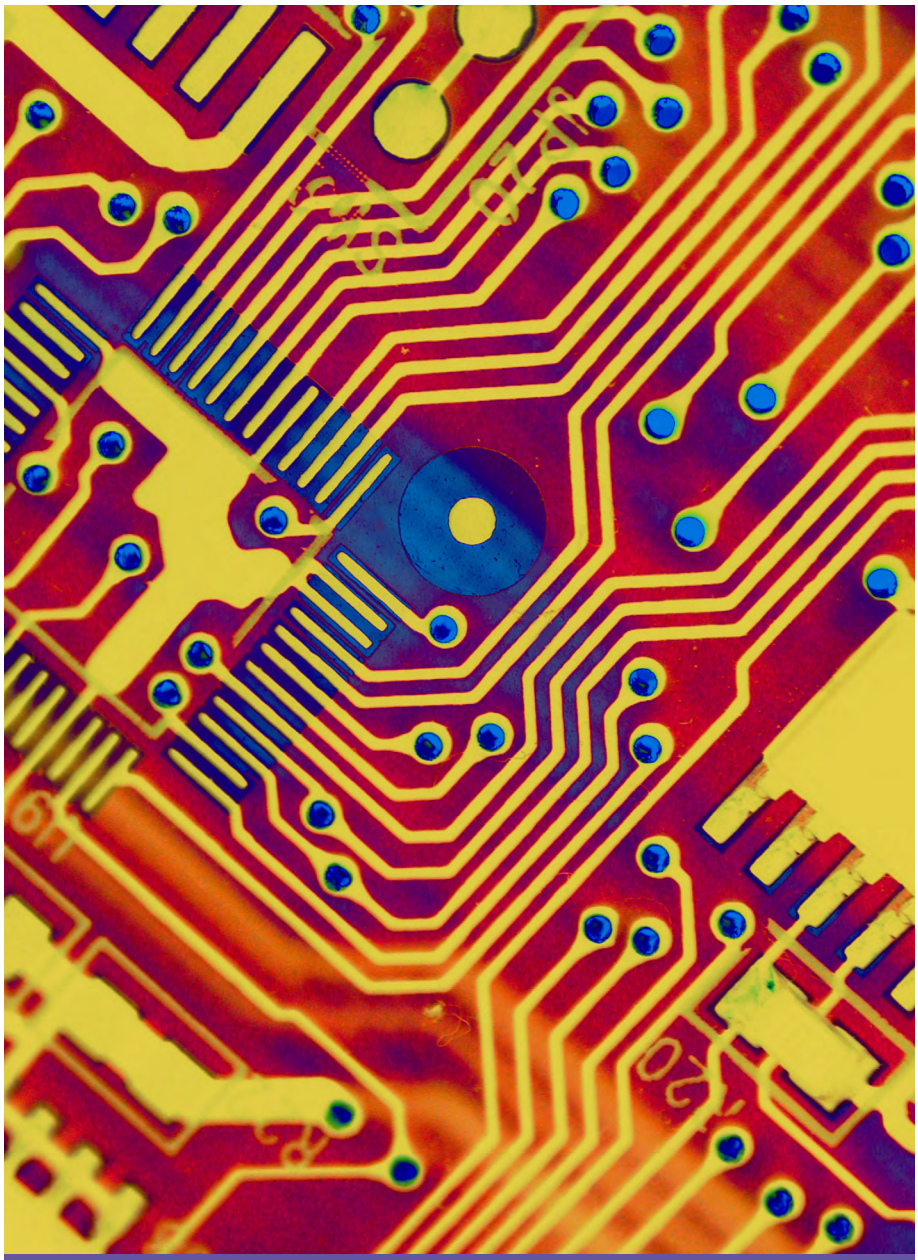

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ALS research in Italy: Deciphering the disease's complexity

AriSLA – The Italian Research Foundation for ALS, outlines strides made in ALS research in Italy over the last 10 years and work still needed to find a cure...

Amyotrophic lateral sclerosis (ALS) is the most common adult onset neurodegenerative disease, characterised by progressive loss of upper and lower motor neurons at the spinal or bulbar level. Motor neurons are the cells responsible for contraction of voluntary muscles which primarily allow voluntary movements, but also other vital functions such as swallowing, speaking and breathing. Their degeneration involves the progressive paralysis of muscles, saving the sensory and in most cases, cognitive functions, blocking the patient in his/her own body.

ALS has a median survival of 2-4 years and the worldwide annual incidence is about 1.9 per 100,000. As recently reported, the number of individuals with ALS is expected to increase by nearly one third from 80,162 in 2015 to 105,693 in 2040. This projection is due to ageing of the population, as those most at risk of developing the disease are between 55-75 years of age. (Nature Communication 2016). This scenario implies the need for common strategies to face the impact that this increase will have on national healthcare systems.

Incredible advances in technology

AriSLA – the Italian Research Foundation for Amyotrophic Lateral Sclerosis, was born in 2009 thanks to the foresight of its founders: AISLA Onlus, the ALS Patients Association, Fondazione Cariplo, Fondazione Telethon and Fondazione Vialli e Mauro per la Ricerca e lo Sport Onlus.

The last 10 years have been, indeed, characterised by an incredible development of new technologies for sequencing and imaging that have allowed us to identify many ALS-associated genes and biomarkers, which are potentially useful for diagnosis and prognosis in relatively cheaper and faster ways, as well as the development of *in vitro* (induced pluripotent stem cells) and *in vivo* tools, that allow us to study what happens prior to symptom onset. This vividness has exponentially

increased our knowledge about the disease and radically changed the mode of operation of the research itself.

Moreover synergies between worldwide charities and associations dealing with the disease and funding research have become stronger and more productive, especially in the clinical and technological areas that have a direct impact on people affected by ALS. This explosion of knowledge has unfortunately raised more questions than answers in the field of ALS.

Lack of disease models and absence of biomarkers hinders progress

ALS is a complex disease with many different heterogeneous pathways playing a role in either its onset or progression. Although most cases of ALS are sporadic (90-95%) with no obvious genetically inherited component, about 5-10% of the cases have a family history (FALS) that occurs due to mutations in specific genetic loci. Starting from the discovery of mutations in the gene, encoding the antioxidant enzyme superoxide dismutase 1 (SOD1) in 1993, more than 50 further potential genes have been identified in association with FALS (Nature 2016).

Even if presentation of familiar and sporadic forms are very similar, the clinical course is highly variable, confirming how the pathogenesis of the disease remains mostly unknown and how complex it is to develop therapeutic approaches that are effective across the ALS spectrum. After half a century of trials and more than 150 different therapeutic agents tested in preclinical models, currently the only available drug treatment is Riluzole, which only modestly increases survival of ALS patients. Potential reasons for the negative results include both the lack of reliable disease models (two-thirds of negative studies were potentially misled by positive studies in SOD1 mice in most cases performed at the presymptomatic stages), and the absence of diagnostic and prognostic biomarkers, which makes the evaluation of the effectiveness of the treatment tested difficult (Lancet Neurol 2014).

ALS research in Italy aims to provide concrete answers

From these premises, the funding model adopted by the AriSLA Foundation aims to support the whole process of scientific research and project development, to act as an interlocutor for the national and international scientific community working on the disease, not only as a funding agency, but particularly as a catalyst of synergies and concrete actions to understand and fight the disease.

Taking advantage of its institutional role and its established network with other international ALS funding charities, AriSLA has invested in all areas of research (basic, translational, clinical and technological), concentrating most of its efforts on the identification of innovative therapeutic solutions and supporting researchers in the creation of a clinical national platform that will help to achieve its main purpose: "Funding the best research to defeat the disease". Through a peer-review selection process, AriSLA wants to be a guarantor of the principles of objectivity, impartiality and scientific merit, using its own Scientific Committee, composed of international, annually renewed experts.

As an example of the results that have been achieved in the last 8 years of research activity, more than €10.6m was invested, with 62 research projects funded and more than 110 research groups supported, to help to produce 163 original publications and lead to the discovery of 5 new ALS causative genes.

AriSLA's effort to increase awareness about the disease is changing "the way to fight against ALS", but there is still an urgent need to implement research investments to develop an effective treatment and improve patients' quality of life.

Exploitation of the new knowledge and new technologies available, together with our investments on translational research, can give us the chance to reach the goal of providing, within the next 10 years, concrete answers to the ALS community. ■

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The role of small heat shock proteins in human diseases

Professor Serena Carra and her collaborators are investigating the important role of small heat shock proteins in age-related neurodegenerative diseases...

Cells are frequently exposed to stress conditions that can damage their proteome, with consequences for their viability. To maintain a healthy proteome and preserve cell function, cells evolved a protein quality control system (PQC). This PQC system consists of molecular chaperones – heat shock proteins (HSPs), small HSPs and co-chaperones – and degradation systems (proteasome and autophagy).

Most HSPs are constitutively expressed; however, HSPs are also induced upon stress to allow cells to respond and adapt, and are therefore referred to as "stress proteins". One of the key functions of chaperones is to cooperate with degradation systems to limit the accumulation of aggregation-prone proteins, a hallmark of many age-related neurodegenerative diseases. Genetic and experimental evidence demonstrates a direct link between imbalances in proteostasis, accumulation of protein-RNA aggregates and amyotrophic lateral sclerosis (ALS), Alzheimer's disease (AD), frontotemporal dementia (FTD), and motor neuropathies. Moreover, mutations in small HSPs and co-chaperones (BAG3) cause neuromuscular diseases.

Due to the increasing average age of the population, the socioeconomic burden of these disorders is incrementing, urging us to find therapies that can delay their onset and alleviate or cure their symptoms. Research aimed at understanding the physiological functions of stress proteins, how they maintain protein-RNA homeostasis, and how their expression is regulated will be essential for the identification of drugs that will ensure a healthy proteome and healthy ageing.

The direct link of small HSPs with human pathologies and their potential as therapeutic targets to cure cardiac and neurological diseases, as well as cancer, was the focus of an international workshop organised by Professor Serena Carra and sponsored by the Cell Stress Society International (Bertinoro, 2016). Worldwide experts in the small HSP field met to discuss two main aspects: first, the importance of supporting interdisciplinary research into the regulation and function of small HSPs; second, the need to understand how mutations in small HSPs lead to disease in humans, therefore providing innovative insight for therapeutic drug design.

Understanding the implication of small HSPs in human disease represents the core of the research program directed by Professor Carra. With the support from the Italian Ministry for Instruction, University and Research, the Italian Ministry of Health, the Italian Foundation for Research on ALS and the Telethon Foundation, and together with her collaborators Professor Simon Alberti (MPI CBG, Dresden), Professor Angelo Poletti and Dr Valeria Crippa (University of Milan), Dr Cristina Cereda (IRCCS Casimiro Mondino) and Dr Jessica Mandrioli (University of Modena), Serena Carra recently discovered a chaperone-mediated quality control pathway called granulostasis that maintains the functionality of stress granules (Ganassi et al., 2016).

Stress granules are ribonucleoprotein complexes induced by cells exposed to stress. While stress granules are highly dynamic in healthy cells, they become dysfunctional and less dynamic in ALS, AD and FTD, ultimately accumulating and contributing to disease. Boosting the HSPB8-BAG3-HSP70 granulostasis complex might help the cells to maintain the functionality of stress granules, thereby delaying disease progression. Carra and her collaborators are synergising their efforts to identify other players of granulostasis and discover drugs that can upregulate the HSPB8-BAG3-HSP70 complex for therapeutic use.

In parallel, the research team also identified mutations in HSP genes linked to rare neuromuscular diseases that are object of current investigations.

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