

A Web-based information system as a tool for the clinical management and support of rare disease patients: the example of amyotrophic lateral sclerosis (ALS)

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BACKGROUND

- a National rare diseases' network exists connecting regional networks (20 Italian regions +2 prov. with health programming competence) thanks to a national law (DM 279/2001)
- Veneto Region (together with Friuli VG, Bolzano, Trento: the "Wide Area") planned a network connecting centres of reference, hospitals, health care local units, Regions, through a unique computerized platform
- ALS is a rare neurodegenerative and rapidly progressive disease
- ALS patients have many health care needs, involving different professionals (neurologist, physiatrist, rehabilitation therapist, dietician, general practitioner, ...)
- a multidisciplinary approach is needed, connecting patients' health issues with social ones
- patients and family members strongly advocate for global care



METHODS

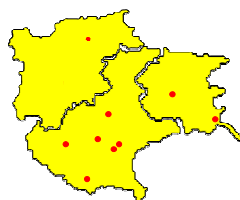
- Data analysis about ALS patients using Veneto Rare Disease Register (4,7 mill inhs) and Hospital Discharge Records
- literature systematic review about best practice and care pathways about ALS (drugs, functional scales, timing, prognostic factors, communication devices, nutrition, ...)
- scheduled meetings of a multidisciplinary technical group of the Wide Area (centres of reference for ALS; health care local units, pharmaceutical services, general practitioners, physiatrists, pneumologists; Coordinating Centre and Register of Rare Diseases of Veneto Region) with the advocacy groups of ALS patients ;
- Consensus about the best practice considering the scientific literature, the clinical experience of the involved professionals and the point-of-view of advocacy groups.

OBJECTIVES



- assessing the state of art of the health care pathways of ALS patients in the Wide Area: number of patients and their characteristics, benefits, best practice, problems
- defining a shared clinical protocol about the care pathways of ALS patients, dealing with the clinical contents (drugs, tracheostomy, AAC, ...), timing, patients' values and wills, coordination of services
- following the principles of equity, best practice, central role of patient and her/his family in the care network, continuity of care
- programming the care pathways of ALS patients, within the existing rare diseases' network of the Wide Area

RESULTS



Centres of reference for ALS in the Wide Area

WHO? WHERE?

Patients with ALS: 438 (55% male, 45% female)
 Prevalence: 8.9/100,000 inhs
 Patients' age: 70 years (mode)
 Median Hospitalization per patient: 2.1 (min-max: 1-14)
 Mean Hospitalization length: 16.9 days (10.5% of hosp. have a length of more than 30 days)

HOW?

The web-based system is the core-tool used to link all professionals involved in the care of ALS patients

- Patient is at the centre of the system
- Every professional, working in the hospital, in the health care local unit, in the Hanicap Commissions, connects to the system, and, through a dedicated page can have information about the patient collected by other professionals and insert information collected by her/himself useful to other professionals.
- The international standardized scale ALSFRS-R, compiled by the neurologist during every visit (every 1-3 months), gives the pece to the clinical pathway of the patients.
- ALSFRS-R scores give alert about what the patient needs according to the protocol, making the involved professionals to heve the neede new information, useful to give benefit or services to the patient

WHAT more?

Drugs, devices, benefits, managment tailored to the specific needs of ALS patients, extra from what is covered by the National Health System

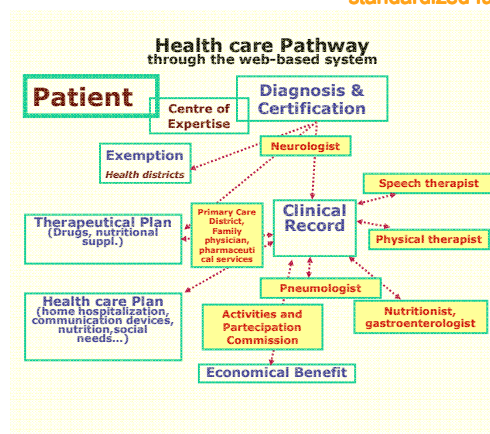
37 essential drugs and dietary supplements in exemption (included off-label, not sold in Italy, galenic products): to heal symptoms (cramps, spasticity, emotional lability, anxiety, sialorrhoea, bronchial secretions, chocking, depression)

Rehabilitation and prothesis-devices useful to the patient according to her/his needs; eye-gaze communication system when needed

Economic and social benefits provided by law (by Activities and Participation Commissions)

Home visits or home hospitalization carried on by local health care and social professionals, but planned by the centres of reference

Scientific literature evidence about ALS patients' care, standardized functional scales, timing are benchmarks



Technologies are useful to serve patients' needs, improving collaboration and communication among different professionals of different insitutions involved in the care process

Shared protocol based on the available evidence as based of a common web-based system answer to the relevant need of equity and translation reserch