

SE L'IMPEGNO NON E' SOLO CLINICO: LE MALATTIE NEUROMUSCOLARI IN EMERGENZA - URGENZA





SMA



**acronimo
malattia
malati
famiglie**



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GENOVA 30 MAG - 1 GIU 2024

Anita Pallara – famiglie SMA







**progetto
cura
strategia
formazione**



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patto di cura





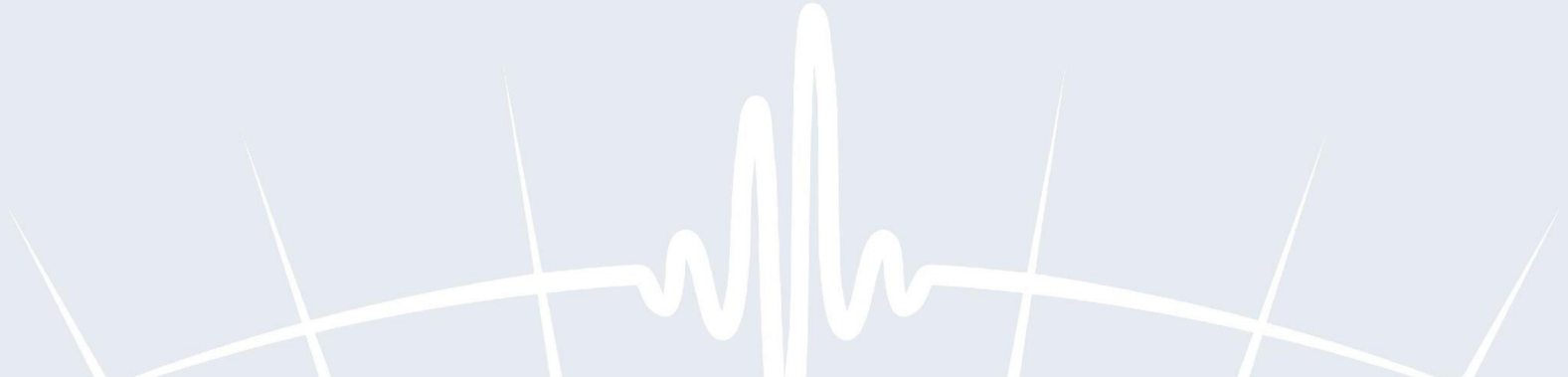
venirsi incontro





umanizzazione delle cure





costruire insieme nuovi percorsi





SPECIAL ARTICLE

Managing emergency and urgent care in spinal muscular atrophy

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The multidisciplinary approach and the active involvement of various professional figures have substantially improved the quality of life of people affected by spinal muscular atrophy (SMA) and given them the possibility of carrying out activities and tasks on a daily basis.

New therapies, which have made it possible to reach survival rates never achieved before, have however opened up new challenges concerning psycho-social needs, the transition into adulthood and the management of health emergencies. These aspects were once considered less but are now increasingly topical and important.

A group of experts was set up to address and propose actions aimed at improving the experience of patients with SMA in Emergency Departments, including representatives of patients and specialists from different backgrounds. The work of the group of experts has resulted in a brief guide with crucial recommendations for the Emergency and Urgent Care setting. The objective is twofold: on the one hand, to improve the ER experience for patients affected by SMA and on the other to make ER staff aware of the clinical and care needs of these patients. ER physicians can play a crucial role in identifying potential risks/exacerbating factors, as well as in taking timely action to effectively avoid the progression of complications in patients with SMA.

cal and care needs of these patients. ER physicians can play a crucial role in identifying potential risks/exacerbating factors, as well as in taking timely action to effectively avoid the progression of complications in patients with SMA.

Current knowledge

The effectiveness of new therapies for SMA management is now widely demonstrated in clinical trials as well as in real life.¹ The results obtained thanks to innovative drugs have opened up the way to addressing the needs of these patients in their daily life that will tend to be increasingly similar to that of the population not affected by SMA.

These needs also include the management of emergencies that require access to the ER. Patients with SMA are particularly vulnerable and subject to respiratory tract infections,² and other manifestations (e.g., pain, gastrointestinal pathologies, fractures) can occur with a certain frequency and require access to the ER for care.³ The hospitals where emergency patients with



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Con i patrocini di **Em SIMEU** **Ministero della Sanità**



GESTIONE DELL'ATROFIA MUSCOLARE SPINALE IN MEDICINA D'EMERGENZA/URGENZA

VADEMECUM



DISTRESS RESPIRATORIO

- NON USARE OSSIGENO**
È controindicato l'uso di ossigeno in assenza di ventilazione non invasiva (NIV). È fondamentale monitorare costantemente la concentrazione di CO₂.
- Garantire la pervietà delle vie aeree
- Garantire la ventilazione manuale:
 - Polmone autoespandibile
 - NIV
 - Presidi in uso (ventilatore portatile, macchina della tosse)
- Definire l'efficienza della ventilazione del paziente (ipercapnia o ipocapnia ipossidemia)
- In caso di **SOSPETTA INFEZIONE DELLE VIE AEREE** (SaO₂ <95% in aria ambiente):
 - Assumere preventivamente una terapia antibiotica empirica.
 - Appena possibile effettuare la **TORACE** e, in assenza di infezione, considerare cause non infettive:
 - PNEUMOTORACE
 - EDEMA POLMONARE CARDIOGENO
 - EMBOLIA POLMONARE
 - EMFISIMA ADIPOSO
- Se il **Torace** non giustifica il quadro clinico di insufficienza respiratoria, effettuare TC torace con mezzo di contrasto per escludere embolia polmonare.



CAREGIVER

IL CAREGIVER DEVE ACCOMPAGNARE IL PAZIENTE (insieme al device e agli aiuti in dotazione al paziente) durante tutto il periodo di permanenza in PS.



INFORMARSI

È sempre necessaria l'informazione sulla presenza di:

- plani condivisi delle cure
- disposizioni anticipate di trattamento



Modulo da De Iaco F, Fontana A, Mercuri EM, et al.: *Managing emergency and urgent care in spinal muscular atrophy*, Italian Journal of Emergency Medicine 2023 Apr 17(2): 4-8
DOI: 10.23756/2523-1285.23.00179-9

Con la partecipazione non esclusiva di



GESTIONE DELLE SECREZIONI

- Usare la macchina della tosse, aspirazioni o altro mezzo/indicazioni, ascoltando anche i suggerimenti del caregiver
- Usare la **MACCHINA DELLA TOSSE** per evitare il rischio di ab ingestis



TRAUMA

- ESCLUDERE SEMPRE IL RISCHIO DI FRATTURE ANCHE NEI CASI MINORI SOSPETTI**
- Farmaci da utilizzare per la gestione del dolore:
 - PARACETAMOLO
 - FANS
 - OPPIOIDI
 - KETAMINA



DISIDRATAZIONE

- Emorragici con glicemia ed elettroliti
- Terapia reidratante tempestiva

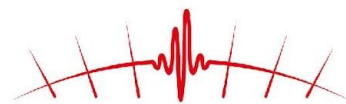


SEDAZIONE

- In caso di necessità di procedure in sedazione coinvolgere un anestesista rianimatore esperto

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***progetto che potremmo definire
utopistico e quasi da sognatori***



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