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ORIGINAL ARTICLE

Spinal muscular atrophy and respiratory failure. How do primary care paediatricians act in a simulated scenario?☆

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Spinal muscular atrophy type 1;
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Paediatrics;
Primary care

Abstract
Introduction: Spinal muscular atrophy type 1 (SMA-1) tends to be fatal in the first year of life if there is no ventilatory support. The decision whether to start such support is an ethical conflict for healthcare professionals.
Materials and methods: A scenario of acute respiratory failure in an infant with SMA-1 has been included in a training programme using advanced simulation for Primary Care Paediatricians (PCP). The performances of 34 groups of 4 paediatricians, who participated in 17 courses, were systematically analysed. Clinical, ethical and communication aspects with parents were evaluated.
Results: The initial technical assistance (administration of oxygen and immediate ventilatory support) was correctly performed by 94% of the teams. However, the PCP had problems in dealing with the ethical aspects of the case. Of the 85% of the teams that raised the ethical conflict with parents, 29% did so on their own initiative, 23% actively excluded them, and only 6% involved them and took their opinion into account in making decisions. Only 11.7% asked about the quality of life of children and 12% about their knowledge of the prognosis of the disease. None explained treatment alternatives, nor tried to contact the paediatrician responsible for the child.

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Conclusions: When faced with a simulated SMA-1 infant with respiratory failure, PCP have difficulties in interacting with the family, and to involve it in the decision making process. Practical training of all paediatricians should include case scenarios with an ethical clinical problem.

PALABRAS CLAVE
Atrofia muscular espinal tipo 1;
Ética asistencial;
Conflicto ético;
Familia;
Comunicación;
Toma de decisiones;
Simulación;
Pediatría;
Atención primaria

Atrofia muscular espinal y fracaso respiratorio. ¿Cómo actúan los pediatras de atención primaria en un escenario simulado?

Resumen
Introducción: La atrofia muscular espinal tipo 1 (AME-1) suele ser mortal en el primer año de vida sin soporte ventilatorio. La decisión de iniciar dicho soporte o no, supone un conflicto ético para los profesionales sanitarios.
Material y métodos: Se incluyó un escenario de fracaso respiratorio agudo en un lactante con AME-1 en un programa de formación mediante simulación avanzada para pediatras de atención primaria (PAP). Se analizaron de forma sistemática las actuaciones de 34 grupos de 4 pediatras que participaron en 17 cursos. Se valoraron los aspectos clínicos, éticos y de comunicación con los padres.
Resultados: La asistencia técnica inicial (administración de oxígeno y soporte ventilatorio inmediato) fue realizada correctamente por el 94% de los equipos. Sin embargo, los PAP tuvieron problemas al abordar los aspectos éticos del caso. Del 85% de los equipos que plantearon el conflicto ético a los padres, lo hizo por iniciativa propia el 29%, el 23% los excluyó de forma activa y solo el 6% los implicaron y tuvieron en cuenta su opinión en la toma de decisiones. Solo el 11,7% preguntó por la calidad de vida del niño y el 12% por su conocimiento del pronóstico de la enfermedad. Ninguno les explicó las alternativas de tratamiento ni trató de contactar con el pediatra de referencia.
Conclusiones: Ante un caso simulado de AME-1, los PAP tienen dificultades para interactuar con la familia e implicarla en la toma de decisiones. La formación práctica de todos los pediatras debería incluir problemas de ética clínica.

Introduction

Spinal muscular atrophy type 1 (SMA-1), also known as Werdnig–Hoffman disease, is a severe neuromuscular disease with an autosomal recessive pattern of inheritance in which degeneration of spinal motor neurons results in progressive and irreversible muscular atrophy. Even though patients can maintain respiratory function in the early months of life, most die within 1 year if they do not receive some type of mechanical ventilatory support.[1,2]

The decision of initiating or withholding this treatment carries important ethical implications and raises considerable doubts in the health professionals involved in the care of these patients in any capacity (neonatologists, neuropaediatricians, intensive care paediatricians, primary care paediatricians, nursing staff, psychologists etc.).[3] A few years ago, the ethical dilemma involved choosing between connecting the patient to an invasive mechanical ventilation system (initially through an endotracheal tube and subsequently by means of tracheotomy) or to limit treatment efforts to comfort measures and terminal sedation. At present, the use of noninvasive ventilation by means of some type of facial interface is a third option.[4,5]

In cases of children with an SMA-1 diagnosis, the decision-making process should include honest information and take into account the natural history of the disease, the opinions of the family, the long-term respiratory and nutritional support options, and care options that limit life-sustaining support. The healthcare team should be interdisciplinary and include the paediatrician in charge and other professionals acting in coordination. At times, the dilemma arises because patients visit the clinic presenting with respiratory failure and neither the family nor the paediatrician know how care should be approached at that juncture.

In recent years, advanced simulation has been used as an interactive method to aid the practical training of health professionals.[6–8] Simulation also allows the analysis of common errors made by participants and the identification of knowledge gaps that need to be remedied.[9–11] While simulation can be used to teach providers how to manage situations that pose ethical dilemmas, we do not know of any studies that have analysed this subject.

Our hypothesis was that primary care paediatricians (PCPs) have difficulty handling cases in which the main clinical issue is of an ethical nature, and the aim of our study was to assess the approach used by PCPs in a case that

poses an ethical dilemma (infant with SMA-1 presenting with respiratory failure) using a simulated environment.

Materials and methods

We generated a scenario of acute respiratory failure in an infant with SMA-1 and included it in the programme of continuing education with advanced simulation for PCPs promoted by the Sociedad Española de Pediatría de Atención Primaria (Spanish Society of Primary Care Paediatrics).[8] In the context of the course curriculum, the educational goal of this particular scenario was to train paediatricians to assess not only the clinical and technical aspects of the case, but also its ethical aspects. The advanced simulation system and educational methodology used have been described in previous publications.[8–10]

Scenario description

The parents of a 2-month-old infant bring him to the PC paediatrics clinic and report a history of increased weakness, pale complexion and fatigue in the past few days. The patient had been hospitalised from birth to 2 weeks prior to the current visit. To make the simulation more realistic, the training includes playing a video of a real patient with SMA-1 with the same clinical presentation. The initial vital signs were the following: oxygen saturation, 85%; respiratory rate, 40 breaths per minute (with shallow breathing); heart rate, 190 beats per minute; and blood pressure, 85/40 mmHg. In the scenario, two people play the roles of the parents of the child, and, if asked, express their disagreement with the treatment approach (one of them in favour of long-term mechanical ventilation and the other in favour of limiting treatment and implementing comfort measures).

Participants

Participation in the study was voluntary, and participants gave oral informed consent both for the recording (audio and video) of their actions during the interactive discussion phase of the advanced simulation sessions, and for the analysis that was the purpose of this study.

Assessment of the scenarios

The videos were evaluated by one of the authors (MBC), who had not participated in the trainings nor knew any of the participants. The analysis of the scenarios took into account the approach of the PCP team involved in each simulation. Of the 49 obtained scenarios, eight were excluded due to technical problems and seven because they included other type of clinicians (hospital paediatricians or paediatrics residents). The final analysis included 34 teams of four PCPs (for a total of 136 PCPs) that participated in 17 trainings. A systematic review was conducted that focused on the hypothesis and objectives of the study, and analysing clinical, ethical and doctor–parent communication aspects, as well as the interventions and procedures used in the management of the patient's respiratory failure.

Statistical analysis

Each health care team was considered a participant in the study. We have expressed the results as the percentage of the total number of participating teams or as mean ± standard deviation.

Results

Most teams (85%) posed the ethical dilemma to the parents, although only 29% did so of their own accord, with the ''parents'' broaching the subject in the rest of the scenarios. Eighty-eight percent did not explore whether the parents knew the diagnosis and the short-and long-term prognosis of the disease.

Of all teams, only 11.7% asked the parents about the previous clinical condition of the child and his quality of life. Only two teams (6%) involved the parents in the decision-making process and took into account their opinion, while 23% actively excluded the family from the process.

None of the teams explained the different treatment options that could be considered for a patient with SMA-1 with respiratory failure to the family, nor attempted to contact the physician in charge of the patient to find out what had been previously discussed or the care plan that had been agreed on for situations like the one presented in the simulated scenario.

In the event of respiratory failure, all teams initiated oxygen therapy after 1.7 ± 0.6 min. When it came to decisions relating to respiratory support, 94% of the teams started ventilation with a self-inflating bag and facial mask, and 62% proceeded to perform orotracheal intubation followed by ventilation with a self-inflating bag.

Discussion

As paediatricians, we should have the necessary skills to adequately manage acute conditions that may emerge in clinical practice from a scientific, technical and also ethical standpoint. While education and clinical experience lay the foundation for these skills, it is difficult to learn how to manage certain situations because they rarely present themselves.[6,8] Advance simulation is a valuable teaching tool that allows the generation of scenarios in a controlled environment to facilitate acquisition of practical skills by health professionals without posing risks to patients.[6,7,11,12] For the most part, advanced medical simulation programmes have focused on technical aspects and in procedures to be performed in emergencies, possibly to address the needs perceived by educators and participants, with a lesser emphasis on non-technical skills.[12]

The aim of our study was to contribute information on the approach of PCPs to a simulated clinical case in which the main concerns were ethical in nature. To this end, we chose to simulate a case of an infant with SMA-1 presenting with respiratory failure, as this disease is a good model of ethical conflict in decision-making relative to respiratory support.[1–4] While there have been attempts to reach an international consensus on the subject, the attitudes of health professionals and families are diverse, so that any of the three possible courses of action (invasive ventilation,

noninvasive ventilation and limitation of treatment) may be taken.[1,2,5] In specific real-life cases, decision-making is not easy and is usually preceded by a process that includes diagnosis and informing the family about the prognosis of the disease and available treatment options, expected quality of life and technical, psychological and social support options.[13,14] In this process, the establishment and communication of a negotiated care plan is one of the objectives that need to be pursued so that every health professional that has to provide care for the patient for any reason knows what needs to be done. Thus, rather than studying the choices made by PCPs in an emergency, our aim was to assess how they approached the ethical challenge of informing the parents, involving them in the decision-making process and implementing the decisions previously agreed on by the healthcare team and the family.

We observed that less than a third of the teams posed the ethical dilemma to the parents of their own accord, with parents otherwise having to bring up the subject with the clinicians. Furthermore, the parents were rarely engaged in the decision-making process either to learn how well informed they were and what their expectations were prior to the clinical deterioration of the child, or to take their opinion into account at the time of the visit. While PCPs may know and provide care for patients with complex conditions, they are not usually primarily responsible for managing the care of these patients nor for making this type of decisions, which are usually the responsibility of specialised colleagues that work in hospital settings. However, we observed that none of the teams asked whether there was a care plan that had been negotiated by the paediatrician in charge and the parents, a piece of information that would be very helpful in decision-making in the event of acute respiratory failure.

On the other hand, when it came to the respiratory care of the patient, all PCP teams administered oxygen therapy correctly and promptly, initiating positive pressure ventilation with self-inflating bags and facial masks followed by endotracheal intubation in 62% of the cases, which demonstrated their ability to quickly deliver appropriate care in instances of acute respiratory failure in infants.

Our study shows that our role as paediatricians requires further theoretical and practical training on the subject of care ethics, shared decision-making and the clinical relationship. The scientific knowledge and technological resources that can be applied to the diagnosis and treatment of our patients keep growing, but it seems that these advances have not been matched by advances in training and resources to address the ethical aspects that arise in the context of the clinical relationship.[15]

There are some limitations to our study. On one hand, there are the limitations resulting from conducting a study in a simulated and controlled environment, as its results may be biased by the methodology and thus cannot be directly extrapolated to clinical practice. On the other hand, while the chosen scenario is considered a good model of ethical conflict, it has particular connotations that prevent the generalisation of the results. In any case, this limitation would apply regardless of which scenario were chosen.

Since the training programme that incorporated the use of advanced simulation was aimed at PCPs, our results cannot be extrapolated to other groups of paediatricians that work in hospital settings or with subspecialty training. In fact, it would be interesting to reproduce the same study in hospitalists (not only paediatricians but also nursing staff) to learn how they approach this conflict. Although the assessment of the actions taken was systematic and involved the repeated reproduction of audio and video recordings, it was conducted by a single researcher, which may have biased the results.

In conclusion, when presented with a simulated case in which the main clinical issue was of an ethical nature, PCPs had difficulty interacting with the family and involving the family in decision-making. The practical training of paediatricians should include scenarios with ethical dilemmas.

Conflicts of interest

The authors have no conflicts of interest to declare.

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